

STATURE, DWARFISM AND INFANTILISM,
IN
MENTAL DEFICIENCY.

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SCOPE OF THE THESIS.

This thesis gives the results of an investigation carried out in a group of 2,103 mentally defective persons in a colony in South-East England.

The main subjects of investigation were:-

(a) the stature of mental defectives as a whole compared with the general population, and normals in their own district.

(b) the types of dwarfism and infantilism likely to be met with in mental deficiency.

(c) the stature and sexual development of special types of mental deficiency.

A table of the types of dwarfism and infantilism met with in mental deficiency is drawn up from the classifications and opinions of various authors published over the past thirty years.

A series of 26 cases, illustrative of the types found, is included.

The index, overleaf, shows the main sections into which the work is divided, and a summary and conclusions will be found at the end.

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DEFINITION OF THE TERMS "DWARFISM" AND "INFANTILISM"

In a survey such as this it is well to begin with a clear definition of the clinical entities which are being described.

The term "dwarf" is rather difficult to define accurately, and one of the earlier descriptions, that of Meige (1895), is "a person of very small size as compared with the average of individuals of the same age and race".

The term "dwarf" is applied both to those who owe their defective stature to local deformity, and to those who, while not deformed, are much below the level of growth for their particular race. Meige's description was added to by Ettore Levi (1910) who inserted the proviso that true dwarfs must possess the somatic characters and functions, particularly sexual, corresponding to their age. This was added so that cases exhibiting infantilism should be excluded. As cases of infantilism may be tall, normal or dwarfed, this addition seems warranted. The psychic, as well as the somatic, immaturity of infantile subjects makes a further division between them and the true dwarfs. Hastings Gilford (1911) also remarks on the difference between true dwarfs and other types. He would make defective growth alone the true criterion of dwarfism and excludes both dwarfism due to local deformity and dwarfism due to "defective development". This latter class he would ally to the infantilisms but qualifies

this statement by admitting that certain individuals are "so far below the mean average stature for their race, and yet in every other respect normal, that they may be termed dwarfs.

In this thesis the classification of Meige is adhered to, with the addition of Levi's proviso to exclude infantilism. This rather narrow definition of the term "dwarf" makes the number of true dwarfs small compared with the number of small people who exhibit infantile characteristics as well.

On a purely statistical basis it is possible to define a dwarf as an individual whose height is less than the mean stature for his race and age by more than three times the standard deviation for that race. In English people this would put the upper limits of dwarfism at about $59\frac{1}{2}$ inches in the case of males and 51 inches in case of females.

Infantilism.

The word "infantilism" was first used by Lasegue (1816-83) in his clinical teaching. In 1871, Lorain first put the word into print in his preface to the thesis of his pupil Faneau de la Cour.

Souques and Chauvet (1913) defined infantilism as a general arrest of the organism with the persistence, in a subject over the age of puberty, of the chief morphological features of in-

fancy,namely,hypoplasia of the genitalia,absence of secondary sex characters,dwarfed stature,delayed union of the epiphyses and a child-like body.

Gardiner Hill (1937) states that the term,"infantilism",should be reserved for a permanent condition in which the physical and psychological attributes of childhood persist into adult life. Smallness of stature,underdevelopment of the musculature,a head large in proportion to the body, comparatively short limbs,lack of function of the genitalia and certain qualities in the mental sphere, must all be present. This gives a clear cut picture of the condition and excludes such entities as eunuchoidism which only fulfils part of the requirements.

The American endocrinologists are not so severe in their description of the condition and Wolf (1939) merely states that infantilism is a condition in which an individual past the age of puberty presents characteristics normally found in one much younger. He is prepared to admit two classes,one of which shows retardation including every part of the body and mind,and another showing only partial retardation of certain physical and mental features.

Gardiner Hill's classification seems best as cases in the second class can only be said to show "certain infantile characteristics".

THE HISTORICAL ASPECT OF DWARFISM.

Dwarfs have appeared in the pages of history from very early times, and records of their appearance and capabilities are to be found in statuary, paintings and stories, both true and fabulous. Many different varieties have been depicted and it is probable that many of them were members of dwarf races who had been captured either by piracy, by the slave trade, or in war.

The dwarf gladiators of Domitian, as typified by statues in the Oppenheim collection of the Bibliothèque Nationale in Paris, appear to have been definitely achondroplastic. Mention is made in the Bible (Leviticus xxi.16-20) of the fact that no dwarf may officiate at the altar. Egyptian statuary often shows dwarf figures and Horus, the child god, has been associated by some with the condition of infantile myxoedema.

In the Roman era, a dwarf was part of the establishment of every important family and Mark Antony, according to Horace, possessed a dwarf called Sisyphus, who was under two feet high. Many of these Roman dwarfs appear to have been achondroplastic. Aesop, of the Fables, is reputed to have been very small and his statuette, in the form of a rickety dwarf, is to be seen in the South Kensington Museum.

During the decline of the Roman Empire dwarfs tended to disappear from courts and large

establishments but in the sixteenth century the fashion reappeared. Catherine of Medici caused marriages to be celebrated between her dwarfs with the object of producing a pygmy race but the result was uniformly barren.

Charles the Ninth of France, in 1572, owned nine dwarfs and the fashion continued up to the eighteenth century. The function of these dwarfs was to provide amusement for the company and many of them were allowed a great deal of licence and freedom of speech. The position of privileged jester no doubt allowed many of them to cloak the inferiority feelings evoked by their small stature under a guise of mordant wit.

In 1713, Princess Natalia, sister of Peter the Great of Russia, married her dwarf Walakoff to the female dwarf of Princess Prascovia Theodorovna. It is not related if there were any children but, subsequently, these marriages were forbidden because of the difficulties and dangers of childbirth when pregnancy resulted.

Celebrated artists have left us many portraits of dwarfs and such examples of Velasquez' work as "Don Antonio, l'Inglese", and "Don Sebastiano de Morra", are world famous.

Dwarfs have been renowned in different spheres of life without necessarily owing their success to the patronage of the great. This is not to be wondered at if we accept the hypothesis that intelligence may have some relationship to brain weight. The brain in certain dwarfs has been shown to weigh

up to one ninth of the total body weight, as opposed to one thirtieth in the normal. Out of many instances the following will give some idea of the diverse ways in which dwarfs have made a name for themselves.

Attila the Hun is quoted by Jornandes in his *Historica Gotthorum* as being of "Forma brevis, lato pectore, capite grandiore". This description appears to put him in the achondroplastic class. Charles Durazzo, king of Naples in 1380, was nicknamed "the Small", and Wladislaus "Cubitalis", king of Poland from 1260 until 1333, although renowned for his daring military strategy, courage and intelligence, earned his name from the translation of a Polish word meaning an ell.

In France, about the years 1605 to 1672, there lived a man called Godeau who was so small that he was always refused in marriage, but he eventually became the Bishop of Grasse. He, however, may have owed something to the patronage of Richelieu.

In this country, during the reign of Charles the First, there was a famous dwarf called Jeffrey Hudson who attended Henrietta Maria, Charles' wife. He was often entrusted with important missions and once travelled to France to superintend the selection of an attendant midwife for the Queen's confinement. His portrait is to be seen both at Hampton Court and at Buckingham Palace, and he is reputed to have been only three feet nine inches high at the age of thirty. He figures prominently in Sir Walter Scott's novel "Peveril of the Peak".

Joseph Boruwlaski, who was born in Poland in 1739, lived most of his life in England. He was a proportional dwarf of true, or "primordial", type and owes his claim to fame by attaining the ripe age of ninety-eight.

In 1843 we find the record of a marriage between two dwarfs which resulted in the birth of fourteen robust and healthy children. These were born, over a period of twenty years, to Robert Skinner and his wife, Judith. This couple was exhibited in London and made a modest fortune. About this time there is mention of a female dwarf called Carrie Akers who was chiefly famous for her enormous obesity. Although she was only two feet ten inches in height her weight reached the phenomenal figure of three hundred and nine pounds!

From the middle to the end of the last century dwarfs were a feature of various exhibitions and to these "show" dwarfs belong such famous personalities as Tom Thumb, General Mite, Commodore Nutt and the Warren sisters. Lavinia, one of the Warren sisters, married Tom Thumb and they were reported to have had one son who was born alive but died in infancy. General and Mrs. Small are reported to have had healthy twins born to them in 1895. Small, who was actually a Welshman called Morris, from Blaenavon, was only thirty five inches in height and his wife was said to be even smaller. Many of these exhibition dwarfs were of the proportionate, primordial type and were possessed of normal intelligence and hardy physique.

At the present time the fashion for exhibiting dwarfs has fallen into abeyance, although there was a dwarf village at the last World Fair in America, and attention is now rather directed to the medico-pathological aspect of the subject. However, from this short description, it may be seen that many of them have led useful, and often famous, lives and have displayed a variety of talents in different fields of life ranging from the sphere of kings and courtiers to the humbler surroundings of the circus.

It is probable that any successes that have come to dwarfs have been confined to certain definite types. These are three in number and comprise the achondroplastic, rickety and true primordial dwarfs. These latter have been termed in English, "ateleiotics", the name given to them by Hastings Gilford. The type is well enough known to be clearly labelled in other languages, such as the German, "Echter Zwergwuchs", and the French "Nanisme Vrai".

Richsbeith and Barrington (1912) give a good description of the historical aspect of dwarfism and many of these facts are to be found in their monograph.

RACIAL AND DEVELOPMENTAL ASPECT OF
DWARFISM AND INFANTILISM.

There are two great classes of dwarfs.

- (1). Racial, or Phylogenetic Dwarfs.
- (2). Individual, or Ontogenetic Dwarfs.

Although this description of dwarfism and infantilism deals mainly with British types one must bear in mind those peoples who are, racially, much smaller in build than our own. The Ghurkas, Japanese, Lapps and Burmese are, phylogenetically, small in stature but this smallness cannot be attributed to defective racial development. Yet, in a British or Scandinavian population many of the members of these races would rank as dwarfs.

Scattered throughout the world we find other, more primitive peoples who are even smaller than the races already mentioned and who represent an earlier, less developed racial stage. Many of these show phylogenetic infantilism as well as dwarfing. Such would be the natives of the uncivilised parts of the Philippine and Andaman Islands, and the pygmy tribes of Central Africa such as the Akkas, Obongos, and Bosjemans (Bushmen.).

As Hastings Gilford states (p.551 et seq.), "Evolutionary infantilism results when the individual and his ancestors have lived for many generations in circumstances calculated to produce developmental stagnation". According to De Quatrefages, Europe, Asia and Africa were, at one time,

peopled by a race of broadheaded, short limbed, swarthy people who were very primitive and whose evolution remained stagnant for thousands of years. These people were overwhelmed, ultimately, by a bigger, stronger and coarser stock whose evolution, due to climatic and other causes, had proceeded along different lines. A tall, long headed, fair and more intelligent type then became masters of the first inhabitants who either became slaves or else had to withdraw to remote and isolated parts in order to exist. The superior type then became completely dominant by intermarrying with, and absorbing, the best of the inferior stock and exterminating the markedly inferior members who became more and more degenerate.

This process still goes on today and those of lower development who have isolated themselves in inhospitable districts still remain of low, phylogenetically infantile, type. The Negritos of Asia and the Negrillos of Africa typify this stagnation level and adults of these races resemble members of the civilised races only at a childish or infantile stage. These are the lower savages described by Ernst Haeckel, and approach nearest to the apes.

Sir Harry Johnston (1902) draws attention to the "infantilo-simian" characteristics of the Congo pygmies who exhibit flat-tipped, flat-bridged noses with wide nostrils, long upper lips, brachycephaly, rounded outlines and persistence of

lanugo. De Quatrefages also states that secondary sex characteristics, in the form of pubic hair, are often lacking although elsewhere on the body hair may be abundant and foetal lanugo persist. According to the same authority, the body of adult Andaman Islanders shows little enlargement around the hips and trochanters, an essentially childish characteristic.

Three points emerge from this account. First, the very low development of these peoples, approaching the ape. Secondly, the fact that development in them is stagnating and almost stationary. Thirdly, that this stagnation is due to the environment as these peoples are always found in the most unfavourable districts of the countries in which they live.

They represent the backwash of the developmental stream and owe the persistence of their type directly to their environment.

THE INDIVIDUAL, OR ONTOGENETIC TYPE.

Naturally, in a race such as the British, the members showing dwarfism, with or without infantilism, will belong to this second, ontogenetic class, and it is these sporadic types which are discussed in this paper.

Nevertheless, the third of the three points made above has a certain importance in the causation of dwarfed and infantilistic types in

our own race and the facts which follow endeavour to show that at least some of the circumstances which are present in the production of phylogenetically infantile races are also to be found amongst communities such as our own and, of course, in Europe as a whole.

If depressing conditions of environment are brought to bear on certain sections of a race of normal development, then stunted stature and infantile characteristics are apt to reappear, should those conditions persist through a series of generations. This has been termed "developmental infantilism" by Hastings Gilford, (p.547.), in contradistinction to the evolutionary type met with in racial dwarfs.

In a people such as ours, these conditions are to be found in the slums of our great industrial cities. The East End of London, from which the great majority of the defectives examined in this investigation come, is a typical locality. An environment consisting of a hand to mouth existence, insufficient food, sordid courts by day and overcrowded rooms by night, produces a physique warped in body and mind, a lowered resistance to disease, a shorter life and early senile change.

It is in this environment that certain individuals, with an inherent tendency to regress, and acted on by a set of predisposing factors which will be enumerated later, revert to an inferior type characterised by a marked diminution of stature infantile proportions, and, if the case be marked,

generalised infantile features.

This depressing environment is not always necessarily wholly external, as Hastings Gilford goes on to point out. (p.557). Some abnormal organ or system may act on the body as a depressive "internal environment" and the body responds to this by a "correlative" form of dwarfing or infantilism, just as it does in response to a depressing external environment, by producing the "adaptive" type.

This postulation of an "adaptive" and a "correlative" form of dwarfing and infantilism is often complicated by the difficulty of deciding between cause and effect and, in many cases, it may be difficult, or impossible, to decide which is the determining factor.

CLASSIFICATIONS
OF
DWARFISM AND INFANTILISM
BY
VARIOUS AUTHORS, WITH TABLES.

The first table overleaf is compiled from Hastings Gilford's "Disorders of Post-Natal Growth and Development", (1911).

Each table is followed by a discussion and comparison with other opinions.

TABLE 1.

1. DWARFISM WITHOUT INFANTILISM.

- (a) Racial, or Phylogenetic Dwarfs.
- (b) Individual, or Ontogenetic Dwarfs.
 - (i) Primary or Essential. e.g. Sexual Ateleiosis.
 - (ii) Secondary, or Symptomatic. e.g. Rickets.

2. DWARFISM WITH INFANTILISM.

- (a) Evolutionary Infantilism.
- (b) Developmental Infantilism, due to:-
 - (i) Environment
 - (ii) Reversion.

This Developmental Infantilism may be:-

- (1) Adaptative, in response to:-
 - (a) Unhealthy surroundings.
 - (b) Intoxications.
- (2) Correlative, in response to:-
 - (a) A degenerated organ
 - (b) An infantile organ.

Either, or both, of these forming a depressive internal environment.

- (c) Asexual Ateleiosis, occurring as a major variation and having nothing to do with environment.
- (d) Brissaud's Myxoedematous Type,
- (e) Lorain's Anangioplastic Type.

Discussion.

Hastings Gilford's table is based mainly on anthropological and biological lines. The following discussion reviews these types and endeavours to select those which are still valid and useful for incorporation in a more modern classification.

The remarks on Racial, or Phylogenetic types made in the previous pages need not be further elaborated.

Individual, or Ontogenetic, Dwarfs fall into two main groups, primary and secondary. The first group, sexual ateleiosis, can be more conveniently discussed later, together with the asexual type.

The Secondary, or Symptomatic Type includes all those conditions which are described by many other authors and which fall readily into place in any classification of dwarfism. This secondary, symptomatic type includes such conditions as achondroplasia, rickets, osteomalacia, skeletal dystrophies, spinal caries and defects due to nervous disease, e.g. poliomyelitis.

Evolutionary Dwarfism and Infantilism due to environment alone is not seen in a community such as ours where there are always other factors at work.

When, however, certain large sections of a civilised race live for generations in unhygienic surroundings and are subject to malnutrition

and the illnesses and intoxications brought about by their unhealthy surroundings , a reversion of type occurs and a Developmental Infantilism will appear throughout succeeding generations. Then, in each generation, the unhealthy environment, acting on a body with an inherent tendency to regress, produces more and more unhealthy types.

In the Adaptative type of Developmental Infantilism the body reacts to a noxious external environment and other harmful external factors to be discussed below.

In the Correlative type of Developmental Infantilism the body reacts to an "internal" environment consisting of a degenerated or infantile part and correlates itself to this.

The harmful external factors which produce adaptative developmental infantilism, hand in hand with the environment, are set out below.

(a) Specific fevers such as typhoid, scarlet fever, diphtheria, influenza and the like. In this group Gilford cites the case of a girl aged nine years who was confined to bed for fourteen days with a severe illness of an unknown nature. From that date growth had practically ceased and at the age of sixteen she looked only half her age. (p.559). One may now suspect in this case that the pituitary had been grossly affected by toxæmia or an acute inflammatory process following the attack of fever.

(b) Tobacco and Alcohol.

There is no direct evidence to show

that tobacco is a causative agent in the production of infantilism. Alcohol was said to be the direct cause in the case of two young children demonstrated by Lanceraux (1896). These two had taken wine from an early age but there was no improvement when this was withheld. As one of the children had had typhoid fever, with accompanying pneumonia and empyema, at an earlier date and the other had an enlarged heart, liver and spleen, it is doubtful if the wine was the sole responsible agent.

Alcohol appears to exert its chief role in the production of infantilism when it is taken to excess by the parents. The importance of its action is, again, rendered suspect when it is remembered that alcoholism is commonest in degenerate and defective stock amongst whom one would naturally expect to find the greatest percentage of stunted and infantile progeny.

(c) Chronic Intestinal Infection.

This plays its part, as we now know, by interfering with the absorption of fats and vitamins from the alimentary system and is responsible for the production of an important infantile type, namely coeliac rickets.

(d) Syphilis causes a generalised toxæmia, or the degeneration of one especial organ or system. In the latter case we may have the production of generalised infantilism of the "correlative type".

Correlative Developmental Infantilism is said by Gilford to occur in response to an

internal noxious environment formed by a diseased or degenerate organ or system, the body adapting itself to this part. Cases of this kind are to be found in the following conditions.

(a) Pott's Disease.

Although it is more usual for tuberculous caries of the spine to lead to dwarfing without infantilism, Marie and Leri (1904) present a case of correlative infantilism due to this cause.

(b) Muscular Dystrophies.

This is to be found in mental defectives who exhibit dwarfing and sexual retardation due to such conditions as pseudo-hypertrophic muscular dystrophy and dystrophia myotonica. A case is shown.

(c) Hepatic and splenic infantilism is mentioned by Hasenclever (1898) who demonstrated three cases of hypertrophic cirrhosis occurring in one family. Each showed defective development. Infantilism associated with splenomegaly, enlarged liver, jaundice and sexual hypoplasia, is described by Hall (1905).

(d) Cardiac:arterial:anangioplastic: mitral infantilism.

Correlative infantilism accompanying a gross circulatory deficiency has been described by many authors and may still be included in tables of the causes of dwarfism and infantilism. It was not seen to any striking degree in this group, but a youth, aged seventeen, fifty two inches in height and sexually retarded, who had stopped growing at

the age of twelve, is described by Gilford (p.569) as owing his infantilism to Raynaud's Disease.

(e) Cerebro-spinal and Neuropathic Conditions.

As Gilford says, (p.571), "Another important series of organs to fail at an early age, and to lead all the other organs along the path to ruin, is the cerebro-spinal. The brain is especially apt to fail early in regressive life and leads to dotage and dementia before the other parts of the body are beginning to degenerate. So is it also with the same system in the infantile and progressive periods of life. In some cases of defective development the most conspicuous feature is the impaired intelligence, or even imbecility, with which it is accompanied. Generally speaking, when we see a case of infantilism in which imbecility is the conspicuous feature we are probably right in terming it "neuropathic"."

This neuropathic type is upheld by authors such as Wolf and Strauch and cases are to be met with in mental deficiency practice. Such conditions as Little's Disease, birth injuries, cerebral vascular catastrophes in infancy, familial demyelinating diseases and, as will be shown, to a less degree, microcephaly and hydrocephaly, may produce stunting and infantile characteristics to a varying extent.

(f) Indeterminate or Mixed Causes.

Under this heading Gilford quotes a case described by Dupré and Pagniez (1902) as "Infantilisme Dégénératif (Type Lorain) compliqué

de Dysthyroïde Pubèrale (Type Brissaud)". This patient was only forty six inches in height, his father and mother were both alcoholics, and he had had whooping cough, scarlet fever and typhoid in infancy, so his condition may have been due to any, or all, of those causes. However it seems likely that he would fit well into the description given in a previous paragraph, of pituitary damage following on a specific fever with a consequent failure of thyreo-tropic hormone.

Ateleiosis is the term introduced in 1902 by Hastings Gilford to describe two main types of dwarfism, the sexual and asexual ateleiotic. He maintained that ateleiosis arose on a different basis from developmental infantilism. It appears as a discontinuous, or major, variation, makes a spontaneous appearance and has a hereditary transmission. All the cases are of pronounced degree and have a strong resemblance to one another. They have the appearance of what Lavater (1797) called "consolidated infancy". Gilford points out that any infantilism is a variation of a regressive character, but in developmental infantilism the tendency to regression is less pronounced than the influence of environment, while in ateleiosis, on the other hand, the tendency to regression is paramount and the influence of environment nil. Ateleiosis is therefore found in every class of society.

The sexual type of ateleiosis would now correspond to the promordial type of Bassoë

(1922) or the hereditary type described by Gardiner Hill (1937). In this type dwarfism alone is present and infantilism is unrepresented except by the small size and proportions of the subject. The famous show dwarfs such as Tom Thumb, Lavinia Warren and Commodore Nutt, were all examples of this sexual type. Their intelligence was normal, their sex life active, as the accounts in the historical section show, and their whole make-up and outlook, on attaining adult life, was mature.

The asexual type, however, is both dwarfed and infantile. His pathogenesis cannot be explained on the same basis as the sexual type, and he must now be placed in a different category. Gilford, although he explained the difference by assuming that the asexual form was "no more than the sexual form in which the reproductive organs had undergone correlative infantilism" (p. 612), suspected that there might be a pituitary basis for the asexual type.

We would now regard them as cases of pituitary infantilism and place them in the endocrine section of any classification.

The only type described by Gilford which was known to have an endocrine basis was the myxoedematous infantilism of Brissaud, so described by him in 1894. We may take it that those cases showed signs of hypothyroidism, and Gilford himself describes the backward state of osseous development, but Tidy (1939) states that the condition shows

many similarities to early Simmond's disease. These cases, then, may fall into either the thyroid or pituitary group but, in any case, they come into the endocrine class.

Grouped with Brissaud's Myxoedematous type Gilford mentions the Anangioplastic type of Lorain, attributing this to delay of, or stoppage of, the development of the vascular system, by various causes. He comments that these two types are the ones recognized by French writers of the period but objects to making them the basis of a classification on the grounds that there are all sorts of intermediate forms between them.

One can no longer explain this type on Gilford's hypothesis and either we must include it with the hypopituitary group, which seems the most likely category into which most of the examples would fit, or else, where the delayed development seems to be directly due to congenital cardiovascular defect, put it into the cardiac group which Gilford already mentions and which is retained in Table IX amongst the mixed types of infantilism.

This survey of Hastings Gilford's classification shows that many of the types which he describes can still be accepted in modern classifications and the main differences are caused by the advance in endocrinological study during the past thirty years.

The endocrinological basis of infantilism amongst mental defectives is often obscure and the importance of environment, both internal and external in Gilford's sense, paramount.

TABLE II

Bassoe (1922) bases his description of dwarfism on a classification by Von Hanseman (1902), as applied to the Meige-Levi definition.

A. Proportionate Dwarfs.

- (1) Primordial, or True Dwarfs.
- (2) Hypophyseal Dwarfs.
- (3) Mixed, or Indeterminate Forms.

B. Disproportionate Forms.

- (1) Achondroplasia.
- (2) Rachitic.
- (3) Cretins.
- (4) Congenital Syphilis.

Discussion.

Proportionate Dwarfs.

The Primordial, or True, Dwarf resembles a normal person seen through the wrong end of a telescope. There are no features, either physical or mental, denoting infantilism. The genitalia are normal in development and function and the osseous system is not backward. The type is familial, occurs slightly more often in males than females and is transmitted by the father. The sella is normal in contour and size. These cases correspond, of course, to the sexual ateleiotics of Gilford.

Hypophyseal Dwarfs were first described by Paltauf in 1891. They are normal in size at birth but soon lag behind and their osseous system shows open epiphyses to a late age so that growth may continue very slowly long after the normal time of stoppage. Accompanying this there is always a greater or less degree of genital and psychic infantilism. Their measurements always remain childish although they may remain alive long enough to show senile changes which often come on early. If this senility is marked enough it may approach Gilford's progeric type. Post mortem examination may not show any gross changes but destructive lesions of the pituitary are common and, as Erdheim (1916) points out, it is usually intrasellar, because an extrasellar lesion interferes with the hypothalamic region and adiposity then becomes a feature as in Fröhlich's Syndrome.

Mixed forms of Proportionate dwarfism were referred to by Sternberg (1919), but they are of doubtful importance when considering proportionate dwarfism alone, as they are usually infantile and most of them are actually disproportionate.

Disproportionat Types.

Those forms of disproportionate dwarfism quoted by Bassoe are well known and agreed on by various authors but the fourth, namely dwarfism due to congenital syphilis, does not appear, from the investigations carried out in this group of defectives, to be a common cause of dwarfing amongst them. Their height, that is the height of mental defectives suffering from congenital syphilis, was found to be actually nearly one inch greater than the average stature for the group.

TABLE III.

A classification of Infantilism with,
or without, accompanying dwarfism, by A. Strauch. (1922)

Group I. Infantilism due to Endocrine Disease.

- (a) with myxoedema and cretinism.
- (b) with absence or underdevelopment of the genitalia.
- (c) with disturbance of the hypophysis.
- (d) with disturbance of other glands.
(Thymus, suprarenal, pancreas, parathyroids.)
- (e) Pluriglandular.

Group II. Dystrophic Infantilism.

- (a) Status Thymolympathicus.
- (b) Angioplastia.
- (c) Valvular lesions, congenital or acquired.
- (d) Parental intoxications such as lead, morphia, mercury, pellagra, malaria, tuberculosis, leprosy and other endemic diseases.
- (e) Congenital Syphilis and extra-uterine early acquired disease such as tuberculosis, typhoid, coeliac disease, nephritis, cirrhosis.
- (f) Traumatic, congenital or inflammatory brain disease.
- (g) Malnutrition and unhealthy surroundings.
- (h) with no detectable cause. Heredity.
Primary defect of the germ plasm.

Discussion.

August Strauch's classification of pure infantilism is based on that of Anton. It agrees largely with the classification of Wolf, another American author, given later. The endocrine section of the table follows generally accepted lines.

It will be seen that Strauch makes the term "dystrophic infantilism" cover all the types of infantilism which are not clearly due to endocrine deficiency, whether the cause be internal or external. This differs from Wolf's view and there is something to be said for the division into separate sections of those cases which owe their infantilism to external factors, such as injuries in the parents, or to internal disease of a debilitating nature.

Such definite conditions as coeliac rickets, and rickets due to renal failure, deserve too, a section to themselves, as they have in Gardiner Hill's table. In Table IX, which is drawn up from a collation of the opinions in the previous tables examined, the term "Cachectic Infantilism" has been used to cover the condition of dystrophic infantilism where the pathology is due to one or more internal factors, and infantilism due to coeliac or renal rickets has been given a separate heading. Wolf's "Effector" group is divided between the Cachectic class, where the less obvious external factors are placed with the other multiple internal group, and an "Other Types" class, where such entities as traumatic brain disease find a place.

It is doubtful if the type of infantilism described by Strauch as due to status thymo-lymphaticus can be said to be a clinical entity in the light of modern work. The general make-up of Mongolian imbeciles is strongly suggestive of thymic dysfunction but infantilism is the exception in these cases. The so-called "thymic" child is rarely so retarded as to be definitely infantile, and there is no apparent reason, if the type does exist, why it should not find a place in the endocrine group which fills the first half of Strauch's table.

TABLE IV.

According to the classification of
E. Apert, (1933).

1. Thyroid Infantilism.
2. Pituitary Infantilism.
3. Suprarenal Infantilism.
4. Renal Dwarfism and Infantilism.
5. Coeliac Disease.
6. Progeria.
7. Hepato-splenic Infantilism.
8. Infantilo-chetivism.
9. Cardiac Infantilism.
10. Gandy's Retrograde Infantilism.

Discussion.

Apert published his well-known book on "The Infantilisms" in 1931. The list given in Table IV is taken from the English translation of the book by R.W.S.Ellis, in 1933.

Altogether, Apert postulates ten types of infantilism, and four of these are so clear cut that they may be accepted without further discussion.

These are:-

- (a) Thyroid Infantilism.
- (b) Pituitary Infantilism.
- (c) Renal Dwarfism and Infantilism.
- (d) Coeliac Infantilism.

Progeria, the sixth type which he mentions, might well now be classed under the pituitary group. Rolleston (1936) compares it to the juvenile form of Simmond's Disease, in the same way that cretinism is a juvenile form of myxoedema. Other authors, Leyton (1937), Beaumont (1937), cite the pituitary as the cause. Goldzieher (1937) describes the untimely senescence of childhood as being due to pituitary failure, but says that chronic adrenal insufficiency may cause a like condition. Leyton and Beaumont refer to it as a questionable factor only, and the balance of opinion supports a pituitary basis.

The third type of Infantilism in Apert's table, suprarenal infantilism, links up with the above. Morlat (1903) ascribed a case of infantilism to adrenal insufficiency in his These de Paris.

Variot and Pironneau (1910) described a case of senile dwarfism due to hypoadrenalism and, more recently, Rabinowitch and Barden (1932) found the medulla of the suprarenal occupied by a lymphoid growth in a case of infantilism. Apert's two cases were females who showed marked dwarfing, retardation of the sexual characteristics and pigmentation of the skin. The first case showed great improvement on the exhibition of powdered adrenal gland by the mouth. Unfortunately, neither case was able to be followed up and Apert, himself, admits that the diagnosis is doubtful. He also quotes Morlat's case. This patient was twenty years old at the time of the investigation and had been under treatment for Addison's disease since the age of twelve. He was sexually infantile, devoid of pubic, facial, and axillary hair, and had small testes and penis. His height was five feet two inches although he had been four feet ten and a quarter inches at the age of twelve. His proportions were adult and his penis, though small, had the appearance of a mature organ on a smaller scale.

Although Apert appears to regard this case as a definite example of adrenal infantilism it would fit readily into the infantilo-chetivistic group. The patient merely ceased to develop further after the onset of a severe illness - which happened to be Addison's Disease - occurring during the growth period. The same picture will occur with any severe protracted illness and the effects are due, not so much to hypoadrenalism as to a generalised involvement of all the endocrines.

No cases have been found in this group where the symptoms are definitely due to hypoadrenalism.

The subject of hepato-splenic infantilism is, like the adrenal type, far from clear, and cases so far described cannot be directly attributed to dysfunction of either, or both, these organs. As Apert says, "If hepatic lesions are very extensive they result in death before there is sufficient retardation of growth to justify the description of infantilism, whereas, if the hepatic lesion is slight, they are compensated for by the processes of regeneration."

The commonest types described are:-

(1) Hypersplenomegalic biliary cirrhosis. described by Gilbert and Fournier, where the height and weight are very much below the average, puberty is aborted or delayed, and adolescents have the appearance of children of twelve or fifteen years. The infantilism is, therefore, not of very great magnitude.

(2) Still's Disease has much in common with the above type except that jaundice, which has been described in the former condition, is not seen in the latter. These patients may be much retarded and one, described by Rosenfeld (1917), who had suffered from the disease from the age of six years, appeared at sixteen to be only half her age. There is said to be no gross retardation of the epiphyseal closure in this type and it might be argued that the severity of a disease such as this, coming on at

an early age, would produce an infantilism on the same lines as the infantilo-cretivistic group. No cases of hepato-splenic type were found in this group.

Cardiac infantilism is generally included in the tables of dwarfism and infantilism but it is agreed that the retardation is variable and, in many cases, slight. Amongst mental defectives where congenital cardiac anomalies are not very rare, the condition may give rise to stunting of growth and moderate infantilism but the type is not of great importance.

In the class to which he gives the name "Infantilo-cretivism", Apert describes a group whose disabilities are due to a general upset of the organism by an outside factor, rather than to an initial disturbance of one or more of the organs on which normal physical and sexual development depend. The word "cretivism" implies literally "puniness", and these cases are, to all intents, comparable to the cachectic infantilism of Gardiner Hill or the idiopathic type of Lorain. The group is included in table ix under the heading "Simple Cachectic Infantilism."

As Apert says, "to enumerate the morbid processes which are liable to cause infantilo-cretivism would be to pass in review all the causes of chronic illness which are liable to affect patients for a sufficiently long time during the growth period." Inanition, avitaminosis, congenital syphilis, tuberculosis, malaria, prolonged suppuration and diabetes are the examples which he gives and

they may be added or compared to the examples which Hastings Gilford gives in his description of adaptive infantilism.

Cases of this type are to be seen often in mental deficiency practice where the above causes are often met with in the family histories.

To conclude Apert's description of infantilism, a less definite type again, first described by Gandy (1906), has to be mentioned. This is known as Gandy's Retrograde Infantilism. Apert, in 1903, was inclined to blame the thyroid but the symptoms might equally be attributed to pituitary failure. The syndrome is characterised by an organic and functional regression of the primary and secondary sex characters towards the infantile state. Regressive atrophy and loss of function of the genital organs takes place and the facial, pubic and axillary hair falls out. Since the patient is past the age of adolescence, the skeletal changes of infantilism cannot take place but there is a certain alteration of the soft tissues and the skin becomes rather bloated and infiltrated. This gives an appearance of age which contrasts with the beardlessness.

The condition resembles Simmond's disease in many respects and may well be due to pituitary failure. The test devised by Fraser and Smith (1941) might prove useful in determining the extent to which this gland was involved. They showed that in the male, the daily excretion of 17-ketosteroids in the urine drops to zero, in Simmond's disease, as

the combined production of the suprarenals (9 mgms) and the testes (5 mgms) is cut off owing to their involution following on pituitary failure.

The syndrome appears to be often incomplete and Allen and Carlyle-Gall (1942) describe a case where there was complete loss of hair, marked depression and loss of libido, without any definite infantilism, the case going on to at least partial recovery.

It must be remembered that, in mental deficiency, one finds cases of regressive infantilism due to such causes as dystrophia myotonica and preceding encephalitis, but the above condition appears to be on a different basis. One imbecile in this series shows complete alopecia of scalp, partial disappearance of secondary hair, ruddy and rather bloated appearance and testicular atrophy with waning libido. The major cause, in his case, seems to be thyroid underfunction but he is not typical of true myxoedema.

TABLE V.

Main types of Dwarfism and Infantilism
described by Rolleston.(1936).

1. Thyroid Dwarfism and Infantilism.
2. Pituitary Dwarfism and Infantilism.
 - (a) Progeria.
 - (b) Simmond's Disease.
 - (c) Babinski-Fröhlich Syndrome.
 - (d) Trauma due to :-
 - Congenital Syphilis.
 - Tuberculosis.
 - Septic Infection.
 - Leukaemia.
 - Neurofibromatosis.
3. Adrenal Infantilism.
4. Retrograde Infantilism of Gandy.

— : —
: —

Discussion.

Adrenal Infantilism appears here again as a fairly definite type. Rolleston quotes the case of Morlat (1903) already referred to, and mentions that Apert, rather hesitatingly as we have seen, also describes the condition. Mott and Hutton (1923) are noted to have described a case of genital hypoplasia which they attributed to adrenal insufficiency. Again we have to bear in mind the close relationship between pituitary and adrenal function.

Rolleston mentions Brissaud's myxoedematous type which the latter described in 1894, and which was attributed by him to a milder form of cretinism. Brissaud described a "moon-like face, flat cheeks, swollen eyelids and thick lips." Rolleston suggests that primary hypopituitarism with secondary thyroid failure may be responsible for these types in certain cases, a finding which is in agreement with Tidy (1939), as we have already seen.

Gandy's Retrograde Type is also mentioned here and a pituitary cause, which Apert also tentatively suggests, put forward as the basis. Rolleston himself suggests that certain specific cases have been due to a pituitary chromophobe adenoma.

Rolleston summarizes the change of views about infantilism which occurred between the years 1902 and 1916 when the role of the pituitary was being taken more and more into account. Gilford (1902), in his article on ateleiosis, quoted

Jonathan Hutchison (1828-1913) to the effect that the cases of ateleiosis exhibiting infantilism would probably be found to be due to some disorder of the pituitary. Levi (1908) stated that Lorain's type, which had hitherto been regarded as due to malnutrition alone, chronic illness or a congenitally defective vascular system (Lanceraux 1893), might be due to pituitary dysfunction. Burnier (1911), Lereboullet et al. (1913), Souques et Chauvet (1913), and Erdheim (1916) all published cases of infantilism for which they pleaded a pituitary basis.

From 1920 onwards the pituitary types were understood more and more fully and Shellin et al. (1934) made a careful study of the subject. Zondek (1935) showed that there was a familial tendency and stated that these types may also show hypothyroidism owing to the failure of the stimulating thyreotropic hormone.

Progeria was first described by Hutchison (1886) and Gilford wrote his description of it in 1904. In 1911, Gilford referred to various other cases and postulated adrenal insufficiency or adrenal tumour as the cause. Apert disagreed with this and showed that primary cortical tumours of the adrenal gave quite the reverse picture. In 1933, Apert attributed progeria to hypertrophy of the thymus but this has not been upheld by later work. Rolleston compares the condition, as already mentioned, to an infantile form of Simmond's disease and a pituitary, rather than an adrenal, insufficiency is upheld as the cause by several other authors.

Simmond's Disease was first described by him in a series of cases published during the 1914-18. These patients showed cretinism, precocious senility, loss of secondary sex characteristics, loss of hair, low metabolic rate, weakness and cachexia associated with infarction of the anterior lobe of the pituitary. This was often associated with sepsis after childbirth, and syphilis. Pituitary cysts and tuberculosis were also shown to be causes by Riecker and Curtis (1932). Certain cases of adult Simmond's disease and Gandy's retrograde type have much in common, in the same way that infantile Simmond's disease and Progeria have points of similarity already noted.

In 1901, Fröhlich, from the Frankl-Hochwart clinic in Vienna, recorded a case of arrested development and obesity in a boy of fourteen years. In the preceding year, Babinski had described a similar case, with a craniopharyngioma of the pituitary, in a paper read before the Société de Neurologie de Paris. Babinski was thus first in the field and although, in this country, the syndrome usually bears Fröhlich's name alone, the two names are often attached to it on the Continent. Other cases were published between the years 1887 and 1906 but the pituitary basis was not clearly recognized, as it had been by these two authors, until the publication, in 1906, of Hervey Cushing. Bartels (1908) introduced the term adiposo-genital dystrophy. The pathology of this type has been fairly clearly

worked out now. Early animal experiments were carried out by Crow et al. (1910) who showed that the removal of portions of the anterior lobe of the pituitary in rats produced adiposity, sexual dystrophy and lethargy. It is now clear that the sexual arrest is due to failure of pituitary function but whether this is due to loss of the hormone elaborated by the basophil cells, or to hypothalamic damage as well, is not known. The upsets of fat, carbohydrate and water metabolism which are often seen are due to hypothalamic pressure and damage by suprasellar growth, often in children, a craniopharyngioma. In adults retrograde symptoms of infantilism may be found with the growth of a chromophobe adenoma of the anterior lobe of the pituitary. Growth is not nearly so much affected as in other pituitary dwarf types and this again points to a hypothalamic, rather than to a strictly limited pituitary cause. Grollman (1941) points out that the tuber cinereum is often chiefly affected and the pituitary body histologically intact.

With special reference to mental deficiency, although cases arising in juveniles are to be found, and these children are often of the "Apache" type with behaviour disorder, more importance can be attached to symptoms akin to Fröhlich's syndrome coming on in adult life and following on previous illnesses such as epidemic encephalitis, basal meningitis, and poliomyelitis. Post encephalitic patients are often obese and show sexual regression and carbohydrate and water metabolism upsets.

One of the patients in this group weighed eighteen and a half stones on admission when she was thirty three years old. She is amenorrhoeic and has signs of mild diabetes insipidus. Her behaviour is very bad and she was unmanageable at home. These symptoms came on in her late 'teens after an attack of encephalitis at the age of ten.

Rolleston gives a few more facts about the role of syphilis in the production of pituitary lesions. Simmonds (1914) has already been noted as describing syphilitic changes in his series of cases. Schmidt (1914) estimated that 47% of cases with syphilis showed pituitary lesions and described interstitial proliferation, miliary gummata and focal necrosis. Skubiszewski (1924) blamed basal syphilis (syphilitic meningitis) as a factor. This condition is one of the causes of hydrocephalic imbecility but in the cases examined here no evidence of gross pituitary deficiency was found. In fact, in view of the relatively large numbers of congenital syphilitics found in mental deficiency, it was surprising how little effect it had in the production of either dwarfism or infantilism amongst those whose Wassermann reactions had been, or were still, positive.

Actinomycosis, septic infection of sinuses leukaemia and neurofibromatosis are also noted by Rolleston to cause infantilism. Von Gierke (1929) described hepato-nephromegalic glycogenic infantilism but admitted that the pituitary was involved. These mixed types would all fall easily into the cachectic infantilism group.

TABLE VI.

H.Gardiner Hill. (1937).

Group A. Simple Dwarfism without Infantilism.

- (1) Racial Types. e.g. Pygmies.
- (2) True Hereditary Dwarfism.(Non-pygmy races.)
- (3) Dwarfism with DEVELOPMENTAL bone disease.
 - (a) Achondroplasia.
 - (b) Chondro-osteo-dystrophy,dyschondroplasia.
 - (c) Osteogenesis imperfecta.
- (4) Dwarfism with ACQUIRED bone disease.
 - (a) Simple rickets. (children)
 - (b) Late rickets. (Osteomalacia in adults)
 - (c) Spinal caries. (Tuberculosis)
 - (d) Spinal deformity. e.g.poliomyelitis.

Group B. Dwarfism with Infantilism.

- (1) Cachectic Infantilism. (synonyms:Lorain's idiopathic:angioplastic:dystrophic:infantilo-chetivism.
- (2) Cachectic Infantilism with Rickets.
 - (a) Coeliac Rickets.
 - (b) Renal Rickets.
- (3) Endocrine Infantilism.
 - (a) Hypopituitary.
 - (b) Hypothyroid.
 - (c) Hypogonadal.
- (4) Other types.

e.g. Cardiac,retrograde,and ? adrenal.

Discussion.

Gardiner Hill (1937) gives a concise and comprehensive classification of dwarfism and infantilism, and Table IX., which is made up of a selection of the established types cited by the authors whose tables have been reviewed, follows the general lines of his table.

He divides the dwarfs into two main groups, those with dwarfing alone, and those with dwarfing plus infantilism. The second group includes cases which may not show much stunting if the cause comes on after adolescence, as in Gandy's retrograde type, or as a result of encephalitis or dystrophy. Many of the indeterminate types found in mental deficiency fall into his Simple Cachectic group and have so been classified in Table IX.

The condition of Mongolian Imbecility has been added to the causes of dwarfism without infantilism, in Table IX.

Certain of the "other types" which Gardiner Hill includes at the end of his table are to be seen in mentally deficient patients and examples of regression after encephalitis, stunting due to cardiac deficiency and a case of (query) retrograde type are shown later and included in Table IX. Adrenal Infantilism has been excluded as no cases were found and, as Gardiner Hill points out, these must be exceedingly rare as the child with bilateral adrenal disease will die young before any clinical diagnosis of dwarfism and infantilism can be made.

TABLE VII.

According to W. Wolf (1939)

- Group A. Endocrine Dwarfism and Infantilism.
- (1) Pituitary.
 - (2) Thyroid.
 - (3) Gonadal.
 - (4) Other glands. (Thymus, suprarenal, pancreas)
 - (5) Pluriglandular.
- Group B. Constitutional Abnormalities with Normal Endocrines.
- (1) Dystrophic Infantilism.
 - (2) Achondroplasia.
 - (3) Renal Infantilism.
- Group C. Affection of certain "Effectors" with a Normal Endocrine and Constitutional Make-up.
- (1) Injury through the parents.
 - (a) by syphilis.
 - (b) by alcohol.
 - (c) by tuberculosis.
 - (2) Unhygienic Conditions.
 - (a) Malnutrition.
 - (b) Illnesses and Intoxications.
 - (3) Traumatic Brain Disease.

Discussion.

Wolf (1939) bases his classification on three points.

- (1) Endocrine Defects.
- (2) Constitutional Abnormalities.
- (3) Affection of certain "Effectors" acting on normal constitution and normal endocrines.

Wolf, and other American endocrinologists, take a broader view of infantilism than British writers. Wolf states that infantilism may be:-

- (1) A condition of retardation of the whole body and mind.
- (2) Retardation of only certain mental and physical features, the development otherwise having proceeded normally.

In mental deficiency, especially in the types due to exogenous factors, one is forced to take this broad view, but the terms, "Regressive Infantilism" or "Infantile characteristics", seem more applicable, the term infantilism being reserved for a widespread condition beginning from birth or a very early age.

Wolf's term, "Dystrophic Infantilism", can be included in the Simple Cachectic Group with multiple disposing factors, and this would also include cases due to injury through the parents and those due to malnutrition and intoxications. The group due to Traumatic Brain Disease is an important one in Mental Deficiency and is included under the Mixed Types in Table IX.

TABLE VIII.A collation of tables i to vii.Dwarfism without InfantilismA. Proportionate Dwarfs.

- | | |
|--------------------------------|-----------------|
| (1) Racial or Phylogenetic. | (Gilford. |
| | (Gardiner Hill |
| (2) Individual or Ontogenetic. | (Gilford. |
| Primary. | (Bassoe. |
| True Hereditary. | (Gardiner Hill. |

B. Disproportionate Dwarfs.

(1) Developmental.

- | | |
|------------------------------|---------------|
| (a) Achondroplasia. | All authors. |
| (b) Chondro-osteo-dystrophy. | Gardiner Hill |
| Osteogenesis Imperfecta. | |

(2) Acquired.

- | | |
|-------------------------------|-----------------|
| (a) Rickety - Simple | (All |
| Osteomalacia) | (authors. |
| (b) Skeletal (Pott's disease) | All authors. |
| (c) Brain trauma. | (Gardiner Hill. |
| | (Wolf. |
| (d) Congenital Syphilis. | Bassoe. |

Dwarfism with Infantilism.A. With a primary endocrine basis.

(a) Pituitary, including:-

1. Asexual ateleiosis. (Gilford)
2. ? Brissaud's Type.
3. Progeria. (Gilford)
4. Simmond's Disease.
5. Babinski-Fröhlich Syndrome.
6. Retrograde of Gandy.

(b) Thyroid.

(c) Gonadal.

(d) ? Other endocrine glands.

(e) Pluriglandular.

(continued overleaf)

TABLE VIII (contd.)B. With a Primary External Environmental Basis.

- (a) Evolutionary.
- (b) Developmental, due to unhygienic surroundings and malnutrition. (Gilford.
(Strauch.
(Wolf.
- (c) External Noxious Agents. (Gilford.
Parental Intoxications. (Strauch.
(Wolf.

C. With a Primary Internal Environmental Basis.

Internal Noxious Agents such as:-

- (1) Degenerated or Infantile Organ, Gilford.
- (2) Congenital Syphilis. (Bassoe
(Strauch.
- (3) Extra-uterine Infantile Disease. Strauch.
- (4) Infantilo-chetivism. Apert.
- (5) Cachectic Infantilism. Gardiner Hill.
- (6) Dystrophic Infantilism (Wolf.
(Strauch.
- (7) Renal Rickets. All
authors.
- (8) Coeliac Rickets. All
authors.
- (9) Cardiac. Lorain's angioplastic, Gilford.
Angioplasia. Strauch.
Acquired Valvular. Strauch.
Cardiac. G-Hill.

D. Other Less Well-defined Types.

- (1) Thymo-lymphatic. Strauch.
- (2) Defective Germ Plasm. Strauch.
- (3) Hepato-splenic. (G-Hill
(Apert.

E. Mixed and Indeterminate Forms.

TABLE IX.Dwarfism and Infantilism in Mental Deficiency.

Group A. Dwarfism without Infantilism.

- (1) True Hereditary Dwarfism.
- (2) Dwarfing due to Developmental Skeletal Disease. e.g. Achondroplasia.
- (3) Mongolism.
- (4) Dwarfing due to Acquired Skeletal Disease.
 - (a) Spinal Caries.
 - (b) Spinal and Limb Deformities due to pre-natal, natal, or post natal brain trauma.
 - (1) Developmental Abnormality.
 - (2) Birth Trauma.
 - (3) Disease Processes.
 - (c) Rickets. (Simple. (children)
 (Osteomalacia. (adults)

Group B. Dwarfism accompanied by Infantilism.

- (1) Simple Cachectic Infantilism with multiple disposing factors.
- (2) Cachectic Infantilism with Rickets.
 - (a) Coeliac Type.
 - (b) Renal Type.
- (3) Endocrine Infantilism.
 - (a) Hypopituitary type.
 - (b) Hypothyroid type.
 - (c) Hypogonadal type.
- (4) Other Types.
 - (a) Traumatic Brain Disease without Limb Deformity.
 - (b) Retrograde.
 - (c) Cardiac.

DWARFISM AND INFANTILISM IN MENTAL DEFICIENCY

(as shown in Table IX.)

Table IX has been drawn up from the classifications reviewed, and in it have been placed those types which may be expected to apply specially to mental deficiency.

Although, in view of the progress which endocrinological research has made in the past two decades, the endocrine aspect of dwarfism and infantilism must necessarily occupy an important place, it is hoped, in agreement with Table IX and the cases included later, to show that a well-marked proportion of the cases in mental deficiency practice have a primary non-endocrine basis, although the glands may ultimately be involved in the regressive process,

Group A of Table IX includes only those types which show dwarfism without accompanying infantilism. In applying the facts from the preceding tables and discussions to mental deficiency it seems unnecessary to divide dwarfism into proportionate and disproportionate types, the former being represented, in Britain, only by the True Hereditary Type. These cases exhibit true dwarfism without any other stigmata except mental deficiency and they are usually feeble-minded. That is to say that they come into the highest grade of mental defect, namely the feeble-minded or moron class. Throughout this paper the terms feeble-minded, imbecile and idiot, are used to describe mentally defective patients

whose intelligence quotients have been found to lie respectively between 50 -70, between 30 -50, and below 30, as measured on the Revised Stanford-Binet Intelligence Scale.(1937). The case of hereditary dwarfing shewn in the examples happens to be a low grade imbecile but this is the exception with this type. She was chosen because there is a well-marked incidence of familial dwarfing in her case.

To this True Hereditary Type, in the first part of Table IX, are added the disproportionate types.

First, Mongolian Imbecility, where the dwarfing tends to resemble that of cretinism or achondroplasia, although not to the same degree. Additional facts about Mongolian stature are given later. Contrary to some opinions, infantilism is not common in Mongols and sexual retardation is found only when hypothyroidism of some severity accompanies the condition. It is uncommon for a Mongolian imbecile to reach the feeble-minded class and many of them are idiots and low grade imbeciles.

Secondly, in the disproportionate class, we include those two groups in which dwarfism is due to either developmental or acquired skeletal disease. The commonest, and more important, members of the developmental group are the achondroplastics. These patients are usually feeble-minded or high grade imbecile in intelligence. Many achondroplastics, of course, are of quite normal intelligence. No cases of chondro-osteo-dystrophy or osteogenesis imperfecta were found in this group, but no doubt osseous



dystrophies are as common amongst defectives as in the general population and, for that reason, they are included in the table.

In acquired skeletal disease we have an important cause of dwarfing in mental deficiency. Spinal caries, rickets and brain trauma are amongst the conditions responsible. Brain trauma, be it noted, gives rise to two main types. The first, which we are referring to now, shows dwarfing alone, due to spinal or limb deformity. The second shows infantilism, with or without dwarfing, accompanied by little or no spinal or limb deformity. Examples of both spinal caries and rickets in dwarfs are given.

Disproportionate Dwarfism due to Congenital Syphilis has not been included in this group although it is cited as a cause by some authorities, such as Bassoe (1922). There are many cases of congenital syphilis in this group but, as the figures show later, dwarfing was not a feature of these cases unless in a few cases where there was involvement of the central nervous system, as in juvenile general paralysis. Such cases may be included in the brain trauma or cachectic groups. All degrees of mental defect can be found in this group. Those with spinal caries, or rickets, grading generally higher in intelligence than those who owe their deformity to brain trauma.

It is interesting to note, however, that the majority of hydrocephalics, many of whom are found at post mortem to have only a thin shell of

cortex above the greatly distended ventricles, have an intelligence quotient which puts them in the upper half of the imbecile group.

Group B. of Table IX gives the main types which exhibit dwarfing accompanied by infantilism. It should be remembered that, in this group, some examples, notably hypogonadism and regressive infantilism, may show but little, or even at times, no, dwarfing.

Simple Cachectic Infantilism, as we have seen in the preceding discussions, includes all those cases which owe their stunted physique and mentality to a diversity of causes which may be endogenous, exogenous, or both. The various terms such as idiopathic, angioplastic, dystrophic and infantilo-cachectic are all grouped under this heading.

The main causes of the types in this cachectic group are summarised in sections B and C of Table VIII, but from these a few types have been separated which rest on a very well defined basis. These are Cachectic Infantilism with either Coeliac or Renal Rickets, Dwarfing and Infantilism due to Traumatic Brain Disease without marked spinal and limb deformity, and Cardiac Dwarfing.

Simple Cachectic Infantilism leads to a very gross retardation in all spheres and the cases in this group were found to be almost without exception, in the idiot grade. Their behaviour, in many cases, corresponds to that of a six months old child. Cases of coeliac and renal rickets, brain trauma and cardiac types, on the other hand, show varying

degrees of intelligence up to high grade imbecile level.

The next section of table ix includes the well known types of primary endocrine infantilism due to pituitary, thyroid and gonadal failure. Other cases where the pathology is more obscure, such as adrenal and pluriglandular types, have been excluded as many of these can be conveniently grouped in the cachectic class. It should be noted in this group that, although infantilism is usually well-marked, stature, which may be very dwarfed in the pituitary class, may be medium in the Fröhlich type, and even above normal in the hypogonadal where, owing to the retardation of epiphyseal closure, pituitary growth hormone is able to act over a longer period than normal. Hypothyroid types, mostly represented by frank cretinism here, are, without exception, markedly dwarfed. Intelligence, in this group, varies with the amount of endocrine failure and the efficacy of replacement therapy. Hypogonadism by itself leads to little intellectual impairment. Pituitary infantilism of the Fröhlich type is often, in mental defectives, accompanied by emotional instability and behaviour disorder. Otherwise the findings are very variable but hypothyroidism, unless treatment is early and adequate, is always accompanied by marked intellectual impairment.

Finally, Table IX presents a mixed group which has been included here because, although the types do not readily fit into the other main groups, yet in the case of mental defect,

they are of sufficient importance not to be excluded entirely.

Brain Trauma is, of course, common among mental defectives and the various lesions which are included under this term often lead to dwarfing and primary, or regressive, infantilism. Such conditions are Little's disease, epidemic encephalitis, meningitis, familial demyelinating diseases, muscular dystrophies and, to a much lesser extent than would be expected, hydrocephaly and microcephaly. Patients in this group show all degrees of mental defect from high grade feeble minded post-encephalitics whose defect lies more in behaviour disorder than in sheer lack of intelligence, to cases of widespread cerebral demyelination who are reduced to the "vegetable" level.

The so-called "cardiac" type is included here because there is no doubt that serious congenital heart lesions do affect growth but the cases seen do not show a great amount of dwarfing or infantilism. If the heart lesion is the sole pathological factor in the case intelligence is not affected except by the lack of education it imposes on these subjects. It may, however, be accompanied by other stigmata, such as Mongolism, in which case the intelligence is much impaired of course.

Retrograde infantilism, as described in the published descriptions, must be rare even in mental deficiency, but it is retained here because it seems to be well established in the literature and actually appears in modified form amongst the

cases in this group.

This group of mixed types could, of course, be lengthened by the inclusion of isolated examples cited by different authorities but, where the pathology or clinical findings are obscure, or where the case may be fitted in under some broader classification already given, it has been thought better to exclude them from this present table.

STATURE IN SPECIAL GROUPS OF MENTAL DEFICIENCY.

Mental Defectives are divided into two main classes.

- 1.Primary Amentia. (endogenous)
- 2.Secondary Amentia. (exogenous)

Eighty per cent of the mentally defective lie in Class 1, and about ninety per cent of this first class is made up of patients suffering from so-called Simple Amentia where the mental defect can be attributed to no single cause, the pathogenesis merely having the label "Germinal" attached to it. The nature of this germinal defect still remains quite unknown although the theories put forward each have a certain amount of evidence in support of them. The average stature of this large group will, of course, tally almost exactly with the average of any large number of defectives.

The remaining ten per cent of Class 1 contains, however, certain important types of mental deficiency in each of which a distinct aetiological factor, or factors, must be at work to produce such a close resemblance between the patients in each group.

These groups are;-

- (1) Mongolian Imbecility.
- (2) Microcephaly.
- (3) Sclerotic Amentia.

Sclerotic Amentia is sub-divided into two types:-

- (a) Nodular. (Epiloia)
- (b) Diffuse. (Macrocephaly)

Other rare types in this group are:-

- (1) Hypertelorism. (No cases seen)
- (2) Naevoid Amentia. (Two cases seen)
- (3) Oxycephaly. (No cases seen)

Of these types, the stature and sexual development of Mongolian imbeciles, microcephalics and sclerotic aments are discussed.

Class 2, Secondary Amentia, accounts for the remaining twenty per cent of mental defectives. The exogenous causes responsible include:-

- (1) Trauma.

Stature in this group will vary with the amount of cerebral neuronie damage and the consequent degree of spinal and limb deformity.

- (2) Infection.

- (a) Meningitis.
- (b) Encephalitis.
- (c) Hydrocephalus.
- (d) Syphilis.

The results of encephalitis, hydrocephalus and syphilis are discussed. The role of meningitis is difficult to determine in most cases.

- (3) Degeneration.

- (a) Amaurotic Family Idiocy.
- (b) Epilepsy.

A case of Amaurotic Family Idiocy is demonstrated and the stature of able-bodied epileptic patients assessed.

(4) Deprivation.

(a) Cretinism, or other endocrine deficiency.

(b) Nutritional deficiency.

(c) Sense deprivation. (blindness etc.)

Sub-groups (a) and (b) naturally show marked dwarfing if the deprivation is present and untreated during the growth period. Cases are demonstrated. Deprivation of sense is not a factor in dwarfing and infantilism unless caused by a debilitating disease occurring during childhood.

Mongolian Imbecility.

The stature is invariably dwarfed.

Twenty female Mongols were examined and the average height was found to be 54.19 inches. None of them was over five feet in height.

Thirty five male Mongols were measured and the average height found to be 56.25 inches. Only five exceeded five feet in height.

Kate Brousseau (1928) gives the average height of 16 female and 22 male Mongols.

Female Mongols - 137.37 cms. (54.78 ins)

Male Mongols - 146 cms. (57.48 ins)

There is especially close agreement between the female figures with those found in this group.

Brousseau states that the body in Mon-

golism is undersized, as in achondroplasia and cretinism, but not to so marked a degree. The shortening of the skeleton of the Mongol is due to decreased length of the long bones rather than to shortening of the vertebral column. The long bones are not thickened, as in the cretin, and do not show the irregularities of growth which characterize cretinism and achondroplasia.

The diagnosis of typical Mongolism is usually apparent even in infancy, the only common error being to confuse the condition with cretinism. Some children, however, show only certain of the Mongolian characteristics and for this reason it is useful to have an examination routine on clearly defined lines as an aid to diagnosis.

Penrose (1933) gives a useful little table which can be applied in the examination of such a case. The incidence of these seven characters is quite significant when Mongolism is contrasted with unselected non-Mongolian defectives. The seven points to look for are:-

- (A) Intelligence Quotient 15-29%.
- (B) Cephalic Index. .83 upwards.
- (C) Epicanthic fold in either eye.
- (D) Conjunctivitis at examination.
- (E) Transverse palmar line, either hand.
- (F) One crease only on minimal digit of either hand.

The table over leaf shows the incidence of these characteristics (frequency per 50 cases)

in two groups of defectives, Mongols and other unselected defectives.

	No.	A.	B.	C.	D.	E.	F.	G.
Mongols.	50.	35.	22.	26.	37.	15.	22.	9.
Unselected								
Cases.	350.	12.9	8.7	1.7	3.6	0.7	1.9	0.1

Actually seven out of those seven points occur in the Mongolian Imbecile demonstrated in this investigation and he was not particularly singled out from the number examined.

Nearly all the ductless glands have been implicated, both severally and collectively, in the search for the aetiology of Mongolism but the thyroid is the one which most often shows signs of malfunction. The hypothyroidism which frequently complicates the condition is presumably coincidental and any improvement in the case after thyroid therapy is due to the treatment of this accompanying deficiency, leaving the Mongolism quite unaffected. The treatment of a Mongolian infant with thyroid, under the misconception that it is a cretin, seems to do more good than harm and seems to lead to some improvement in the general intelligence.

Dysfunction of the thymus has been suggested as a cause by several authors. (Barnes N.P. 1923). Many Mongols show signs akin to those of thymic dysfunction, notably, reduced resistance to infection, low blood pressure, acrocyanosis, lymphocytosis, and angio-neurotic reactions. In

those Mongols who have had radiological examinations of the chest during the past four years in this hospital, there have been no findings suggestive of thymic enlargement.

So far as the gonads are concerned, there is no evidence, in the series of cases examined here, that infantilism is at all common amongst Mongols. None of the male cases has undescended testicles and the genitalia and secondary sex characters are normally developed. Amongst the females, although menstruation is sometimes tardy and irregular, this is probably due rather to the accompanying hypothyroidism than to any effect wrought by the condition of Mongolism itself.

In Mongolism, therefore, we find a condition which, even amongst other defectives, is characterized by diminutive stature. Compared with the general population we find that male adult Mongols are shorter than normal males by more than twelve inches, and that female Mongols are ten inches less in height than normal adult females.

Taking Karl Pearson's normal of 67.68 inches and subtracting therefrom three times the standard deviation (2.7), we arrive at the figure of 59.78 inches. Any person below this stature may, statistically speaking, be said to lie right outside the limits of deviation in the population examined, hence, by definition, they are dwarfs. Male Mongols in this series are actually, on an average, 3.53 inches less than this upper level of dwarfism.

Microcephaly.

Tredgold (1937) states that microcephalics, as a class, are the smallest of defectives and that few of them grow to more than five feet. As many of the patients in this group do not exceed five feet in height, the male average being 62.3 inches, one would not expect a microcephalic, who has a severe organic disability, to reach even this moderate height. Actually, out of the four adult male microcephalics found here, two were exactly five feet high and the average for the four was 57.75 inches, a height which exceeds the average male Mongol by one and a half inches.

Penrose (1933) states that microcephalics are often quite well developed although usually smaller than the normal.

There was only one true microcephalic adult female and her height was 59 inches which is actually 0.04 inches more than the average for the females in the group. All these cases, as will be seen from the example shown, have the true microcephalic head where the decreased cranial capacity is chiefly in the upper part of the skull, the face appearing relatively broad in contrast. One must distinguish the condition from that of nanocephaly in which the head is symmetrically decreased in all its measurements. Many dwarfs have a cranial capacity no greater, or very little greater, than a true microcephalic but have normal intelligence. These are to be considered as examples of nanocephaly

and show none of the stigmata of the true microcephalic type. The microcephalic head may have a circumference greater than 17 inches, the usual criterion, but it must always fulfil certain conditions to be classed as such. These are:-

1. Face appearing relatively large with narrow lower half and receding chin.

2. Long and narrow heads, i.e. the length of the head is nearer normal than any other measurement.

3. Consequent rather low cephalic index.

To sum up the subject of stature in microcephaly we may say that it is less than the average defective's but appreciably greater than the average Mongol's. Accompanying infantilism is not usual and was not seen in any of the cases examined here.

Sclerotic Amentia.

(a) Nodular Type.

This may be called epiloia, tuberous sclerosis or adenoma sebaceum. The three cardinal signs are mental deficiency, adenoma sebaceum of face or body and epilepsy. The clinical and pathological findings are fully discussed by Critchley and Earl (1932) and here only the findings with regard to stature and sexual development will be discussed. Critchley and Earl state that these cases are noticeably undersized for their years without, however, showing any infantilism or absence

of secondary sexual features. Adiposo-genital dystrophy has been described in one case by Lazar(1932). Eleven cases were examined in this group. It was not found possible to assess the male cases as they were mainly adolescent, but six female cases had an average height of 57.75 inches which is 1.21 inches less than the mean for the group. Epiloia does, then, lead to stunting of stature as compared to the normal but not significantly as compared with fellow defectives. None of the cases seen here were infantile and the males tended, if anything, to macrogenitosomia, but without other hyper-sexual features.

(b) Diffuse Type.

From the case demonstrated it will be seen that there are few points of similarity between these two conditions clinically. Epiloiacs are introverted, spiteful and psychotic in a primitive kind of way. Diffuse sclerotics are cheerful, slow and simple and usually quite well extroverted. Proliferation of neuroglia takes place throughout the whole brain leading to enlargement and great increase of weight of that organ and the skull which surrounds it. Tredgold (1937) states that the majority are undersized. As only one case was found in this group whose stature was slightly above the group average, no conclusions could be drawn. This patient shows no signs of infantilism and reached puberty at the normal age, but shows some girdle obesity which has been remarked on by others.

No cases of hypertelorism or oxycephaly were found in this group. Two cases of naevoid amentia, with facial naevus and venous angioma of the meninges showing calcification on radiological examination, were seen. Both of these patients, one male and one female, are feeble-minded and epileptic. This supports Tredgold's assertion that the condition does not lead necessarily to mental deficiency, although contralateral paralysis and epilepsy of Jacksonian type are common.

No anomalies of stature or sexual development were apparent in these cases.

Extrinsic Causes of Mental Defect.

(1) Traumatic lesions of the central nervous system are very common amongst defectives. The causes are legion but an important one is birth trauma. This usually means intra-cranial haemorrhage, often of venous type, and arising, in order of frequency, in the dura, the subarachnoid space, the cerebral substance and the ventricles. There is usually great difficulty in separating these cases into traumatic or developmental groups but the general impression gained from examination here was that the developmental type of paralysis either quadriplegic or diplegic, leads to a much greater degree of stunting than the traumatic and is quite often accompanied by infantilism. The case of cachectic infantilism demonstrated shows to what

degree this dwarfing may attain when quadriparesis is a complicating factor in a childhood beset by constant ill-health in other ways.

A significant percentage of these traumatic cases suffers from lesions of the basal ganglia producing the condition of choreo-athetosis. Here only the extra-pyramidal tracts are involved and the "paralysis" takes the form of a disabling difficulty with the initiation of muscular movement, including speech, which is explosive in type, accompanied by coarse choreic movements of the limbs and head. In those cases where the lesions are entirely extra-pyramidal there seems to be no interference with growth or sex to any degree, but when there is an accompanying involvement of the pyramidal system, and this is often the case, then the stature depends, as aforesaid, on the amount of spinal and limb deformity which results.

(2) Epidemic Encephalitis.

Although the numbers are small and the conclusions drawn, therefore, not very significant, attention is directed to the conditions following on the above illness.

Five adult male cases and six adult females showing clinical signs of a post-encephalitic nature were examined. It was found that no great change in height from the rest of the group could be demonstrated. The males were actually 3.2 inches above, and the females 2.86 inches below the mean stature for the group.

No signs of Infantilism were found amongst the males apart from the loss of libido which may accompany the general apathy found in certain of these cases. Amongst the females, on the other hand, none menstruated regularly, four not at all, and two irregularly. This condition would therefore appear to cause a definite increase in menstrual irregularities perhaps through basal brain changes affecting the pituitary or hypothalamic areas.

A case is shown which exhibits a fair degree of stunting accompanied by primary amenorrhoea following on a verified attack of encephalitis in infancy but this case is again the exception rather than the rule and its inclusion in the female group is responsible for pulling down the height of the female group to a much greater extent than would be found in a larger series.

From the above facts we may deduce tentatively that epidemic encephalitis does not lead, as a general rule, to dwarfing but may lead to sexual retardation, regression or irregularity.

(3) Hydrocephalus.

Tredgold (1937) states that the majority of hydrocephalics are undersized but show no stigmata of degeneration. It was not found that hydrocephalics in this group were much smaller than other patients with a severe organic disability. The average height of six adult hydrocephalic female defectives was 56.59 inches. This is 2.37 in-

ches less than the mean for females in the group. Four male hydrocephalics averaged 61.13 inches in height which is 1.17 inches below the male mean. So it may be seen that in spite of their severe handicap which leads in all the marked cases to paralysis of the lower ,if not all four,limbs,these patients do not show any great growth defect when compared with their fellows although undersized in comparison with normals.

Again,although one might expect pituitary or hypothalamic damage to follow on the conditions found at the base of the brain in hydrocephalics who have had intra-cranial birth haemorrhage,basal meningitis of acute or syphilitic origin,and tumour formation,yet infantilism is not generally evident,some few only showing retardation of puberty.

(4) Syphilis.

The findings amongst patients in this group suffering from congenital syphilis have been referred to previously. This prior statement is amplified below. Bassoe (1922) includes this condition amongst the causes of dwarfism but this is not substantiated by the findings here. Where the cerebro-spinal fluid has been serologically negative it has not been found that the stature of defectives was in any way impaired in comparison with their fellows who did not suffer from the disease.

On the other hand,a case is shown which

shows gross retardation with a positive Wassermann reaction in the cerebro-spinal fluid and a paretic Lange colloidal gold curve. These cases are, however, the exceptions.

Amongst the 2103 cases in this investigation, 62 females and 26 males suffer from congenital syphilis verified by a positive blood reaction at one time or another. The Wassermann reaction tends, in these cases, to become negative as the patient grows up. This has led to much discrepancy in the results reported by investigators on the percentage of sufferers from congenital syphilis amongst defectives. Stewart (1925) reported 19.75 per cent in 800 cases and Hall (1926) found only 1.15 per cent in all the boys admitted to the Rome State School for Defectives (America) during the period 1915-1925. Penrose (1933) shows that the incidence is higher when the group tested is small.

The figures for this group, given above, show that the incidence is 4.1 per cent. This agrees fairly well with the figures for groups of above a thousand quoted by both Tredgold and Penrose.

The average height of the males was found to be 63.15 inches, that is 0.85 inches above the average male stature and well outside the dwarf "standard". The average of the 62 females was 59.9 inches which is 0.94 inch above the female average.

There does not seem to be any justification for including congenital syphilis amongst

the causes of dwarfing in mental defectives, and on these findings it seems likely that real dwarfism amongst normals is rarely due to this condition.

Where the central nervous system remains unaffected infantilism does not accompany congenital syphilis. 66 per cent of the 62 females examined menstruated regularly and this compares creditably with the unaffected remainder of whom 73 per cent have regular periods.

For the reasons set out above it has not been thought justifiable to include congenital syphilis as a cause of dwarfism in the table prepared to show the varieties met with in mental deficiency practice.

(5) Degeneration.

(a) Amaurotic Family Idiocy will give the same picture as any other chronic debilitating disease arising during the growth period and, if the patient survives so long, will lead to a condition of cachectic infantilism. This condition does not always arise in infancy and juvenile types are not uncommon. There have been three cases, all verified by post-mortem, in this hospital. All these cases were delayed in onset until childhood (8-10 years) and the final condition of helpless, vegetative dependence to which they all attained in their 'teens was not accompanied by the gross stunting and sexual dysplasia one would expect to find if the condition had arisen earlier. One of these

cases is included amongst those demonstrated but unfortunately she died before a photograph could be taken. The other two cases died before this investigation began and accurate figures of stature are not available but they were not dwarfed and the other female menstruated irregularly.

(b) Epilepsy.

Out of 2103 defectives in this group 604, or 28.7 per cent, were epileptic. Penrose (1933) gives the average as 20 per cent. Tredgold (1937) puts it at 37 per cent amongst defectives who have no paralyses or other gross cerebral lesion. Crippled patients are said to show a far greater incidence amounting to 75 per cent, and Tredgold states that the incidence increases with the degree of mental deficiency. The figure here seems to be an average one and may be heightened by the fact that a predominantly low grade institution will contain a greater percentage of cripples. In order to assess the effect, if any, of epilepsy on stature, it is necessary to select those who suffer from what, for want of a better term, is described as idiopathic epilepsy.

103 epileptic males who had no gross nervous lesions or other defect except primary amentia were measured. The Intelligence Quotients varied from below thirty to a maximum of seventy, the majority being imbeciles. The mean stature was found to be 63.76 inches, 1.46 inches above the

male mean stature.

Epilepsy, then, as a concomitant of simple mental deficiency, does not lead to any significant stature changes and certainly not to dwarfism. Infantilism or sexual retardation is no commoner than in any comparable group of otherwise able-bodied people.

STATURE OF MENTAL DEFECTIVES
COMPARED WITH OTHER CLASSES OF THE POPULATION

The majority of the patients examined here, as has been said, come from the poor homes and slum districts of Greater London where one would expect find those conditions conducive to stunting of mind and stature.

Tredgold (1937) does not give exact figures in assessing the height of mental defectives as a distinct group, nor does he compare the heights in different parts of the country or in different classes of society. He states that giantism is the exception in mental deficiency, as it is in the general population, and that the average height of mental defectives is "several inches" below that of normal people.

From the table which he quotes we see that lunatics and aments are the smallest of the three classes mentioned only in the oldest age group. Criminals are actually smaller in younger groups.

TABLE X.

		Average height at age of:-			
		No.measured. 20/25 25/35 35/45 45/55 yrs.			
General Population.	5669.	67.5"	67.9"	67.9"	67.9"
Criminals.	2315.	65.2"	65.6"	65.7"	65.8"
Lunatics & Aments.	1409.	{----- 65.7" -----}			

In order to compare the stature of the defectives examined here with that of the general population in the same part of the country, that is London and district, Table XI, prepared by Pearson and Lee (1903), is shown below. This gives the average stature of a predominantly middle class type in the London district. It will be seen from Table XIII that the defectives in this group are quite markedly smaller than Pearson's and Lee's average. The male defective is found to be 5.8 inches less than the normal and the female defective 3.52 inches smaller than the normal.

TABLE XI.

K. Pearson and A. Lee. (1903).

	Mean Stature. (inches)	Standard Deviation.	Coefficient of Variation.
Fathers.	67.68	2.7	3.99
Mothers.	62.48	2.39	3.83
Sons.	68.65	2.71	3.95
Daughters.	63.87	2.61	4.09

-----"-----
"

Although the figures in Tables XI and XIII are taken from normal and mentally deficient subjects in the same part of the country the difference in height will tend to be too great owing to the fact that the figures of Pearson and Lee were assessed on a predominantly middle-class population.

With a view to putting this in better perspective Table XII, in which Clement Dukes (1905)

points out the difference between the favoured and artisan classes, is appended.

TABLE XII.

	Age.	Mean Stature.
Artisan Class.	23-30 yrs.	66.5 ins.
Favoured Class. (Public School, Naval and Military Academy, University.)	25-30 yrs.	69.0 ins.

" "

Table XII still shows a distinct bias in favour of the artisan class - actually 4.2 inches - and adds to the evidence that the living conditions and nutrition, in the even more depressed classes from which the defectives here come, play an important part in the causation of further physical and mental dwarfing.

TABLE XIII.

2,103 Mental Defectives of both sexes, predominantly imbecile. (I.Q. below 50.) Working-class. London area.

Male cases. Total Number. - 1008.

Number measured.	Mean stature.	Standard Deviation.	Coefficient of variation	Standard error.
237.	62.3"	3.07	4.9	$\pm 2(.2)$

Female cases. Total Number - 1095.

Number measured.	Mean stature.	Standard deviation.	Coefficient of variation.	Standard error.
254.	58.96"	3.16	5.19	$\pm 2(0.19)$

Finally, Table XIV gives figures, compiled by Goddard (1912), which show the average height of normal people and mental defectives in the United States of America. This table shows that while there is little difference between American and English normals there is a distinct bias in favour of American defectives. This is as much as 2.3 inches for males, but only 0.14 inch in females. It is probable that Goddard's group contains a greater proportion of morons (feeble minded) than this group and as is shewn later these are definitely taller. Environment and nutrition are also factors, both probably being superior in Goddard's group.

TABLE XIV.

	Age.	No. Measured.	Mean Stature.
Male Defectives	21.	184.	64.7 ins.
Male Normals	21.	-	67.6 ins.
Male Defectives	31-70	637.	64.6 ins.
Male Normals.	31-70	-	67.7 ins.
Female Defectives.	21.	160	60.5 ins.
Female Normals	21.	-	63 ins.
Female Defectives	31-70	960.	60.1 ins.
Female Normals	31-70	-	62.7 ins.

RELATIONSHIP OF INTELLIGENCE TO STATURE
IN MENTAL DEFECTIVES.

In order to find out if there was any relationship between intelligence and stature in mental defectives two separate investigations were carried out.

In the first, 127 adult, able-bodied, female defectives were examined and their intelligence quotients (I.Q.'s) were assessed on the Stanford - Binet Scale. As idiots are not testable on this scale, this group consists only of imbeciles and feeble-minded patients, whose I.Q.'s range from 25% up to 70%. Idiots are presumed to be those whose I.Q.'s are below 25%.

A table was drawn up, (Table XV), in which these 127 defectives are separated into nine I.Q. groups each with a range of 5 points. The percentage of patients in each of these I.Q. groups who were above the average height for the total group, and the percentage of patients who were below the average for the total group, was found, and these percentages are shown in the table.

From analysis of the table it can be seen that:-

(1) 89% of those with an I.Q. lying between 25% and 30% are below the average height of the total group.

(2) 73% of those with an I.Q. lying

We can thus see, from points one and two above, that there is a significant difference in height when we compare the highest, feeble-minded group (I.Q. 65-70%) with the lowest grades of the imbecile group (I.Q. 25-30%)

The stature of the imbecile class (I.Q. 25-50%) corresponds almost exactly to the average height of female defectives as a whole but those whose I.Q. rises above 50% to 70% show a significant increase in stature.

There is thus no constant rise of stature in proportion to the rise of I.Q. but there is a significant difference between the statures of the imbecile and feeble-minded groups. As this difference is most marked at the extremes of the table we are led on to enquire, in the second part of the investigation, whether the difference is even greater when the lowest group of all defectives, the idiots, (I.Q. below 25%), is compared with the feeble-minded class.

The results are shown in Table XVI but before we examine them an important point must be considered. In these tables we are comparing only able-bodied patients. It must be remembered that it is amongst the lower grades of mental deficiency that we find the greatest numbers of patients with physical stigmata. If we examine all those patients who may be classed as idiots and low grade imbeciles we find that many are dwarfed merely by extensive crippling and deformity.

In assessing the relationship of stature to intelligence in these two groups, it is, therefore, necessary to compare samples from each class which consist only of patients who can fairly be said to be able-bodied. In the feeble-minded group, this includes the greater percentage of their numbers but, in the idiot group, by far the greater part consists of patients showing a diversity of crippling disabilities which would lead to stunting of themselves irrespective of the intelligence quotient.

On investigation, 34 female idiots were found who were both adult and able-bodied. 29 male idiots were also measured. A random series of 34 feeble-minded adult women was also taken for comparison with the idiots and 47 able-bodied adult male morons were contrasted with the male idiots.

TABLE XVI.

Group.	Number.	Mean Stature.
Female Idiots.	34.	56.7 ins.
Female Feeble-minded.	34.	60.9 ins.
Male Idiots.	29.	59.9 ins.
Male Feeble-minded.	47.	65.8 ins.

-----"-----
"

From this we see that:-

(1) Female idiots are 2.26 inches below the mean stature for the group.

(2) Feeble-minded females are 1.94 inches above the mean stature for the group.

(3) Male idiots are 2.4 inches below the mean stature for the group.

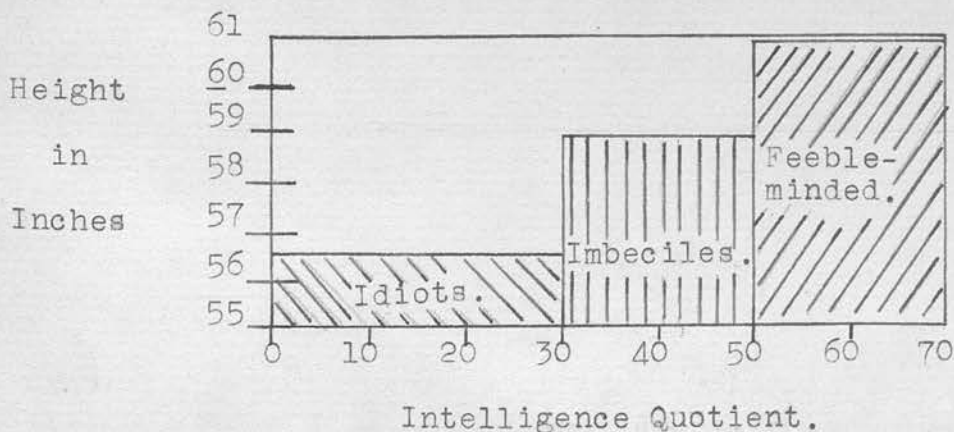
and (4) Feeble-minded males are 4.9 inches above the mean stature for the group.

Table XVI also shows that feeble minded males are only 0.7 inch shorter than the average height given by Clement Dukes for the artisan class of the London district.

As we have said, to include all the idiots in the group would make their average stature very appreciably less but, setting all those aside whose dwarfing is due to pure physical disability or marked endocrine dysplasia, and making intelligence per se the sole criterion amongst groups of able-bodied patients, we find that stature declines in each intellectual grade from the feeble-minded through the imbecile class to the idiot level.

Table XVII, below, shows this graphically for the three groups.

TABLE XVII.



The relative heights of feeble-minded, imbecile and idiot patients.



Case No.1. Example of Hereditary Type of Dwarfism.

Sex. Female. Grade. Idiot. Age 26.

Family History.

Father, aged 78. Health good. Ht. 64 ins. Wt. 142 lb.

Mother, aged 58. Health good. Ht. 59 ins. Wt. 116 lb.

Siblings. Patient is the fifth of six pregnancies.

The second child, a male, died in infancy. The rest are all normal. Nearly all the members of the family on both sides are small. "Around five feet to five feet four".

Personal History.

Birth was normal. She has never talked. Walked at one year. Teething normal. No illnesses of note.

Habits. A quiet, timid girl who never speaks but hums to herself. Doubly incontinent.

Physical Examination.

General Appearance. Tiny but well proportioned except for a slight dorsal kyphosis.

Height. 49 ins. Weight 63 lb. Vertex-Symphysis 24 in.

Symphysis-Soles 25 ins. Span 50 ins.

Fat. Well covered. Posture - slight kyphosis.

Skin. Smooth and fine. Teeth. Fairly regular and well calcified. Hair. Healthy. Rather hirsute, with luxuriant axillary and pubic hair. None on face.

Eyes. Healthy, and fundi normal. Neck. No thyroid or cervical glands palpable. Breasts. Well formed and proportionate. Genitalia normal. Menses began at the age of fourteen and a half years. Systems otherwise normal.

Laboratory Investigations.

Urine. No abnormal constituents.

Blood Pressure. 110/70.

Wassermann Reaction - Negative.

Red Blood Cells. 3,800,00 per c.m. Hb. 72% (Sahli)

Colour Index. .95.

Serum Calcium 10.5 mgms %. Inorganic Phosphate 3.45 mgms %.

Glucose Tolerance Curve. "Lag" type. No glycosuria.

Radiological Examination. Sella turcica normal.

Wrists. Epiphyses fused, carpals massed and calcification good.

Summary.

A small, well proportioned woman of twenty six years. There is a well marked family history of dwarfing but none of mental deficiency or endocrine defect. The trunk-limb proportions resemble normal, the whole body merely being on a smaller scale. The only sign of endocrine defect is in the menstrual history where the periods, having begun at the normal time, have since become irregular. The blood chemistry is not abnormal except for a "lag" type of glucose tolerance curve. There has never been any glycosuria during her ten years stay in hospital.

Although this patient is an idiot, the majority of cases of this type have normal intelligence.



Case No 2. Example of Achondroplasia. Sex.Female.
Age 50. Grade. Imbecile.

Family History.

None is available. She was admitted to hospital at the age of ten.An illegitimate,unwanted child. Parents unknown. Siblings.Patient says she has none.

Illnesses.

During her forty years in hospital she has had no serious illnesses of any kind.

She is employed in the needle-room doing simple mending.

Habits and Personality.

She is a neat and tidy woman. Extremely shy and introverted. Rarely speaks unless spoken to and is very docile and industrious.

Physical Examination.

General Appearance. A tiny woman with the typical signs of achondroplasia.

Weight. 76 lb. Height 47.5 ins.

Vertex-Symphysis 28 ins. Symphysis-Soles 19.5 ins.

Span 46 ins. Upper arm 9 ins. Lower arm 11.5 ins.

Thigh 9 ins. Leg 10.5 ins.

Fat. Well covered. Posture erect. Skin supple and smooth. Hair healthy. No hypertrichosis. Pubic and axillary hair rather sparse. Teeth are regular, not overcrowded and, though the enamel is ridged, they are less carious than those of defectives of similar age. Eyes, some blepharitis. Fundi normal.

Neck. No thyroid or cervical glands palpable.

Breasts. Menopausal, atrophic changes. No mastitis.

Genitalia. Normal for age. Menses were regular up to the age of forty-five when they ceased.

Other findings normal.

Laboratory Findings.

B.P. 110/70. R.B.C.'s 4,200,000. Hb. 80%. C.I. 0.95.

Urine. No abnormal constituents.

Wassermann reaction negative.

Summary

A case of achondroplasia showing the usual proportions and exhibiting no accompanying endocrine dysplasia. A quiet, introverted imbecile.



Case No.3. Example of Mongolian Imbecility.

Sex male. Age 29. Grade Idiot.

Family History.

Father died, aged 36, of tuberculosis.

Mother had a paralytic stroke, aged 58.

Paternal grandfather died of tuberculosis.

Maternal grandmother died, aged 32, cause unknown.

Maternal grandfather died in a mental hospital.

Mother's aunt and uncle still in mental hospital.

Aunt is epileptic. A sister has chorea.

Siblings. Patient is the sixth of eight children.

The second died of fits, aged 2. Remainder healthy.

Personal History.

Birth. Seven months baby. Mother 29 years old.

Walked aged 5. Talked aged 5. (Very gruffly)

Teething normal. Not clean in habits until his
'teens.

Illnesses.

Measles, whooping cough, chicken-pox. Chronic ear discharge. Operation for blocked tear duct.

Habits. He must be classed as an idiot as his Intelligence Quotient is below thirty. He is still faulty in his habits and his speech is almost incomprehensible.

Physical Examination.

General Appearance. A typical Mongol who shows all the main stigmata including a long body length compared with the legs.

Weight. 92 lb. Height 55 ins. Vertex-Symphysis 30 ins. Symphysis-Soles 25 ins. Span 58 ins. Cephalic Index 0.84.

Fat is normal. Abdomen a little protuberant.

Posture erect with some lordosis. Skin rather harsh and dry. Hair. Wiry and strong. Axillary hair normal and pubis well covered. Mouth. Tongue large and fissured, teeth large and carious. Speech is very gruff and indistinct. Eyes show slight right internal strabismus, blepharitis, conjunctivitis and slight corneal opacity in right eye due to trauma by ingrowing eyelashes. Fundi normal. Epicanthi.

Neck. No thyroid or cervical gland enlargement.

Genitalia. Normal in all respects. Both testes are descended and of normal size and consistency.

No other gross abnormalities found.

Laboratory Findings.

B.P. 122/84.

Urine. No abnormal constituents.

Wassermann reaction negative.

Summary.

A Mongolian imbecile, aged 29 years, who is four feet seven inches in height. He shows a marked lengthening of the trunk in comparison with the limbs. The lower measurement is five inches shorter than the upper. The span is greater than the total height and the build is different from the achondroplastic.

The genitalia are quite mature, as is found consistently throughout the group of adult Mongols here. The only "infantilistic" signs are the short stature and the childish mentality.

This patient shows all of the seven diagnostic points in Penrose's table.

A. Intelligence Quotient 15-29%.	Plus.
B. Cephalic Index. 0.84	Plus.
C. Epicanthic fold.	Plus.
D. Fissured tongue.	Plus.
E. Conjunctivitis.	Plus.
F. Transverse palmar line.	Plus.
G. One crease on minimal digit.	<u>Plus.</u>
	<u>7.</u>



Case No.4. Example of Dwarfing due to Spinal Caries.

Sex.Male. Grade Imbecile. Age 34.

Family History.

Father, aged 63, health good. Height 66 ins. Wt. 154 lb.

Mother dead. Cause - Insanity. Paternal grandfather was a mental defective.

Siblings. Patient was the last of four pregnancies.

One brother and two sisters are healthy.

No other relatives are of small stature.

Personal History.

Birth was normal. Mother depressed during pregnancy.

Illnesses. Measles, chicken-pox, tuberculous disease of the spine.

Habits. Inoffensive, dull. Does no work.

Physical Examination.

General Appearance. A small, bald-headed, stooping man with marked disproportion between body and limbs.

Height 51 ins. Vertex-Symphysis 23 ins. Symphysis-Soles 28 ins. Span 58 ins. Weight 94 lb.

Fat. Rather lean but with fat abdomen.

Posture. Marked kypho-scoliosis. Limbs straight.

Skin clear and supple. Teeth well spaced, carious.

Hair. Bald. Secondary sex hair plentiful.

Eyes healthy. Fundi normal. No enlargement of the thyroid or cervical glands.

Respiratory System. The chest is very cramped and narrow. Poor excursion. There is no pathological activity.

Genitalia are normal. Testes are descended and no "cragginess" detected.

Laboratory Findings.

B.P. 160/110. Urine contains trace of albumin.

Urea Range (15 grams urea) 3.2 gms% - 0.7 gms%.

Wassermann reaction negative. X-ray shows collapse with angulation of 7-10th dorsal vertebrae.

Summary.

Dwarfing due to spinal caries with no limb involvement. No endocrine dysplasia. Some impairment of renal function.



Case No.5. Example of Dwarfing due to Spinal and
Limb Deformity.

Sex.Male. Grade.Feeble minded. Age 24 yrs.

Family History.

Father aged 45 yrs.Healthy.

Mother,aged 22 at patient's birth. Died of pneumonia next year.

Siblings. Patient is the fourth of family of four.
The others are all healthy. No other members of the family are of small stature.

Personal History.

Birth was normal. He talked at the usual age but did not walk until he was about ten years old. Even then he was only able to shuffle with bent knees.

Illnesses. Measles in infancy. Operation for "glands" aged 3. Legs tenotomised at age of fourteen.

Physical Examination.

General Appearance. A pleasant looking boy who is unable to stand upright by himself and needs a stick or other support. Spinal curvature and flexion contracture of legs evident.

Height. 51 ins (standing). Symphysis-Vertex 27 ins.

Symphysis-Soles 31 ins. Actual height 58 ins.

Span 62 ins. Weight 104 lb.

Fat normal. Hair glossy. Pubic and axillary plentiful.

Posture very stooped and contracted. Skin sallow and rather greasy. Teeth large, regular, yellow but well calcified. Eyes healthy. Fundi normal.

No thyroid palpable, or other cervical glands.

Respiratory System. The chest is cramped and moves badly. Protruding sternum and marked spinal curve. There are no lung lesions detectable.

Central Nervous System. Pupils are equal, circular and react to light and accommodation. All four limbs are spastic but the legs are much more affected than the arms. The patient is capable of co-ordinated arm movements and is employed in mat-making. The legs are much contracted and slightly wasted. All the deep reflexes are uniformly increased on each side, especially the legs. Abdominal and cremasteric present. Plantars extensor on both sides. Other systems are normal. Genitalia are mature.

Laboratory findings.

Urine. No abnormal constituents. Wassermann reaction is negative. B.P. 130/85. Blood Cholesterol 160mgms%. Slightly increased sugar tolerance curve. X-ray shows spinal curvature. No angulation or collapse.

Summary.

This case shows dwarfing due solely to spinal and limb deformity. The contracture of the limbs and the bending of the spine produce a standing height of only 51 inches. The actual body length is 58 ins. The lesion is a congenital spastic quadriparesis of developmental type. When he was seven his thyroid was said to be enlarged and he still has a vaguely "cretinoid" appearance. This juvenile myxoedematous condition seems to have greatly abated and he does not need any thyroid therapy at present. The sugar tolerance is slightly increased and the blood cholesterol within normal limits.

There is no gross angulation of the spine, as in the preceding case, the whole merely being curved in an antero-posterior plane especially in the upper dorsal region. There is no scoliosis. Due probably to disuse there is some osteoporosis of the long bones.



Case No.6. Example of Hydrocephaly. Sex female.
Grade.Feeble minded. Age 15.

Family History.

Father aged 49.Health good.Ht.70 ins. Wt. 159 lb.

Mother.Died aged 49. Cause - cancer. Dyspeptic.

Height 49 ins. Weight 119 lb.

Grandfather intemperate. Grandmother insane.

Siblings. Patient is the last child of five.

Second brother died of tuberculous meningitis,aged
2½ yrs. The rest,brothers,are healthy.

Birth was difficult. Instruments used. One arm was
fractured. She was seen to be hydrocephalic from
the first. Never walked. Incontinent. Talked and
teethed normally. No illnesses of note

Habits. A gentle,docile girl who has been taught
to do simple needlework and knitting.

Physical Examination.

General Appearance. A tiny,crippled and misshapen hydrocephalic girl.

Height 49 ins. Vertex-Symphysis 23 ins. Symphysis-Soles 26 ins. Half span,on less contracted side, is 26 ins. Circumference of head $26\frac{1}{2}$ ins.

Fat,Fairly well covered. Posture.Cannot stand.

Chest misshapen and right arm and leg contracted.

Skin white and fine. Hair fine and silky.

Eyes.Healthy.Fundi are not abnormal although the discs look pale. Vision is quite good.

Neck.No thyroid or cervical glands palpable.

Breasts are immature. Genitalia are sparsely covered with hair and axillary hair is appearing.

Menses have not yet appeared.

Respiratory System.The chest is almost fixed and the sternum protrudes to the left. There is dorsal scoliosis.. The lungs are healthy.

Cardio-vascular System.She has a permanent pulse rate of about 100 per min. Both heart sounds are clear and closed. B.P. 110/74.

Central Nervous System. Pupils are equal,circular, and react to light and accomodation.There is a spastic quadriparesis affecting mainly the right side. All deep reflexes are unduly exaggerated. Bilateral extensor plantar reflexes are present.

Laboratory findings.

Urine.No abnormal constituents.

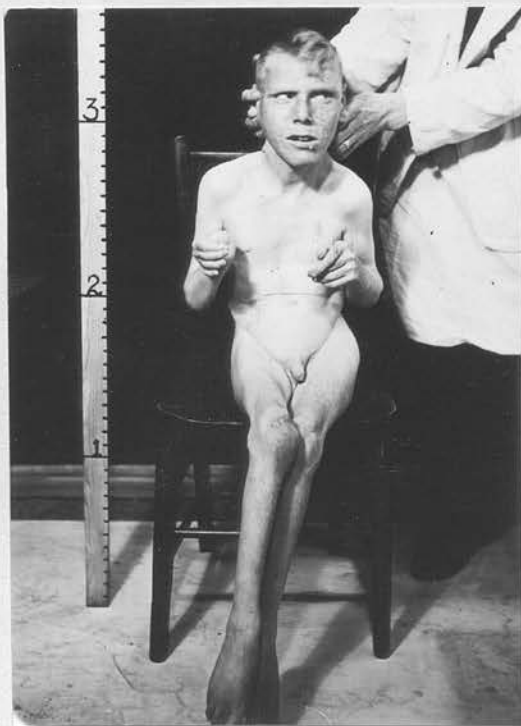
Wassermann reaction negative.

Summary.

This little hydrocephalic girl is shown, mainly, to bring out the gross deformity which can result from this condition with, however, the preservation of a much greater amount of intelligence than one would naturally expect. She is able to carry on a conversation and has been taught to knit in spite of her almost useless right arm.

The secondary sex characters are delayed but beginning to appear at the age of fifteen. Her height is definitely dwarfed but this stunting may be increased by hereditary factors. Her mother was only four feet one inch in height. Growth has not yet ceased. She is much the most crippled of the hydrocephalics in this group but her intelligence compares favourably with those in whom only slight leg spasticity is present.

It is not the general rule for hydrocephalics to show infantilism and their stature, though smaller than that of their able-bodied fellow defectives, does not usually fall within the limits of dwarfism.



Case No.7. Example of Infantilism accompanying
Congenital Syphilis and Little's Disease.

Sex Male. Age 17. Grade.Idiot.

Family History.

The patient's parents could not be examined.

The maternal grandfather died of tuberculosis.

The great-uncle of the patient is a mental defective.

Personal History.

Patient is the third child of three. The others,
when last heard of,were said to be healthy. At
birth the cord was twisted round the neck and he
was very quiet for some hours after birth. The head
was retracted on the second day and he took the
bottle badly. He seemed to hold himself continually
stiff. He became epileptic at the age of seventeen.

Physical Examination.

General Appearance. A small, stunted, crippled boy who, as he lies in bed, has the look and mannerisms of a child of eight. He cannot speak although he cries loudly.

Weight 49 lb. Height 46 ins. Vertex-Symphysis 23 ins. Symphysis-Soles 23 ins. Span 24 ins.

He is rather thin. Posture. A contracted, spastic, quadriplegic, totally bedridden.

Skin is freckled on exposed surfaces. Smooth and fine.

Hair is strong and wiry on head. None in axillae and only a few sparse hairs on pubis.

Teeth are small and carious. Secondary dentition.

Eyes. Healthy. Fundi show patches of pigmentation in choroid round the discs. The thyroid is not palpable.

Central Nervous System. The pupils are equal but give only a sluggish reaction to light. There is slight, right internal strabismus. Cranial nerves otherwise unaffected. The deep reflexes are uniformly exaggerated except in the contracted right arm. The abdominal and cremasteric reflexes are absent. Plantar reflexes bilaterally extensor. There is no loss of pain sensibility, as far as can be determined in an idiot.

Genitalia are very immature and small. Right testis is undescended. Left testis of moderate size. He has no seminal emissions.

Laboratory Findings.

Wassermann Reaction in blood. Positive.

C.S.F. W.R. plus. Lymphocytes 3 per c.mm.

Lange Colloidal Gold curve. 55544431000.

Summary.

The boy shown here is seventeen years of age and is greatly stunted and sexually immature. He comes of defective stock and suffers from congenital syphilis. The blood Wassermann reaction was positive from an early age but the cerebro-spinal fluid was negative until re-examined at the age of seventeen. It had probably been positive for some years before this but was not re-examined until then. When he was sixteen years old he began to have generalised epileptic seizures for the first time in his life.

He shows both dwarfing and infantile characteristics and these are due to the added damage done to the nervous system by generalised syphilitic changes superimposed on extensive crippling due to agenesis or cerebral birth injury.

He compares unfavourably, both physically and sexually with patients suffering from either congenital syphilis or spastic quadriplegia alone.



Case No.8. Example of Rickets giving rise to Dwarfism,with accompanying acromegaly.

Sex male. Age 30. Grade. Idiot.

Family History.

Father aged 34 yrs.,mother aged 30 yrs.,at time of patient's birth.

No history of insanity,paralysis or alcoholism in the family.

Siblings. Three brothers and two sisters whose physical condition was said to be poor.

No other family history available.

The patient was always physically and mentally retarded from birth.Never talked.Stood up at the age of five. Walked at nine. Was able to walk by himself in his 'teens. Previously needed help.

Physical Examination.

General Appearance. A tiny, misshapen, pot-bellied dwarf with large head, jutting jaw, and hands and feet too big for the rest of his body.

Height 40 ins. Vertex-Symphysis 20 ins. Symphysis-Soles. 20 ins. Span 48 ins. Weight 56 lb.

Fat. Very little. Rather wasted. Posture is bent forward at hips. No real kyphosis.

Teeth are fairly regular but poorly calcified and carious. Skin shows marked acrocyanosis. Hair is thick and healthy. Pubic and axillary hair normal. Eyes healthy, fundi normal. No thyroid is palpable and the cervical glands are not enlarged.

The chest is very short and the ribs are splayed out by the swollen abdomen. Breath sounds are faint, vesicular and unaccompanied by crepitations. Heart rate 100 per min. Accentuated second sound at pulmonary area. B.P. 128/85.

Alimentary System. "Pot belly." Small umbilical hernia. Large mass palpable continuous with liver dullness and extending almost to pelvic brim.

This was subsequently found to be faecal.

Genitalia normal in size. Both testes descended.

Urine is normal. Generalised spasticity of limbs but walks quite well.

Laboratory Findings.

Wassermann Reaction negative. X-ray shows de-calcification of long bones. Epiphyses closed. Carpals massed. Tufting of phalanges of hands and hooking of thumb phalanx. No other investigations owing to premature death of patient.

Summary.

As will be seen from the photograph, the patient was a stunted, little creature whose head appeared too big for his body, and whose jaw was large and prognathous. Both the hands and the feet, although smaller than normal, (compare the nurse's hand in picture) were much larger than one would have expected. Owing to the premature death of this patient investigation was rather sparse. He was suddenly taken ill one evening, became enormously distended, and, in spite of digital removal of faeces and enemata combined with carbachol injections, he died of paralytic ileus and cardio-respiratory embarrassment within a few hours.

There was little bossing of the skull, the teeth though carious and badly calcified were regular, the sternum was prominent and costo-chondral beading was seen. The ends of the long bones - radius, ulna and femur - showed enlargement and, radiologically, osteoporosis especially above the epiphyseal lines. The epiphyses were all fused. There was well marked tufting of the distal phalanges and hooking of the distal phalanx of thumb. This latter is said by Werner (1942) to be pathognomic of acromegaly. Genu valgum of slight degree was present.

Apart from the fact that there was not the slightest sign of infantilism present in this case, the appearances and findings, such as they are, would apply to the coeliac type of rickets.

It is probable that he was a rickety baby but the changes found at the time when he was examined may well be due to diminished absorption of calcium from the bowel during years of intestinal stasis. At post mortem the stomach was empty, the small intestine distended and full of faeces in the lower three feet, the large intestine greatly distended and full of faeces from end to end except for the transverse colon which was blown up with gas. The sigmoid colon was especially distended and the walls were greatly thickened.

The intra-abdominal tension was so great that death took place due to sheer cardio-respiratory embarrassment and at the post mortem, which was delayed owing to legal difficulties, almost an explosive rush of gas ensued when the peritoneal cavity was opened. No abnormalities were found in the endocrines macroscopically. The sella was not enlarged and the acromegaly appears to be distinct from the rest of the pathology. The patient appears to have been a mixture of childhood rickets with superimposed adult rickets due to defective absorption. After epiphyseal closure the anterior pituitary became overactive. As the gonads were normal there is no reason to suppose that epiphyseal closure was delayed beyond the normal time, so that the patient, unable to increase in stature, gradually acquired well marked acromegaly over a period of about ten years.



Case No.9. Example of Rickets accompanying Mongolism.
Sex.Male. Grade.Idiot. Age 25.

Family History.

Father,aged66,healthy but undersized and also appears to have suffered from rickets.

Mother died,aged 47,after abdominal operation.

Siblings..The patient is the third of four.

Elder brother and sister normal. The younger brother is a cripple (? cause) in hospital.

No history of mental defect or insanity in family.

Personal History.

Never talked clearly.Did not walk until five years.

Measles,diphtheria twice (tracheotomised once), and pneumonia in infancy.

Habits. An idiot who can do nothing for himself.

Physical Examination.

General Appearance. An undersized Mongolian imbecile with a stooping carriage and bowed legs. Height 49 ins. Vertex-Symphysis 26 ins. Symphysis-Soles 23 ins. Span 54 ins. Weight 74½ lb. Fat. Rather thin. Skin dry and rough. Hair is coarse and strong. Pubic and axillary plentiful. Posture. Stooping with protruding sternum, some lordosis, bent knees and bowing of legs. Mouth. Large fissured tongue which tends to protrude. Teeth consist of a few carious stumps. Eyes. Blepharitis. Neck shows old tracheotomy scar. No thyroid or cervical glands palpable. Chest is ill-formed with a protruding sternum and beaded costo-chondral junctions. Expansion is practically nil. Lungs are clear. Abdomen shows a small umbilical hernia. No enlargement of liver or spleen. Genitalia are normal. Both testes descended. Other abnormalities. Polydactyly.

Laboratory Findings.

B.P. 95/70. Urine. No abnormal constituents. Wassermann Reaction Negative. Glucose Tolerance Curve normal. Serum Calcium 9.1 mgms %. Inorganic phosphate 4.4 mgms %. X-ray shows double terminal phalanges on right thumb. Left amputated (? same condition.) Thickening and bowing of the tibiae with ragged epiphyseal lines still visible at the lower ends of radius and ulna and upper end of ulna.

Summary.

A case of Mongolian Imbecility with rickets as a complicating factor. There is a family history of the latter condition but no incidence of mental deficiency. The patient shows six out of seven of the diagnostic points mentioned in case three. He shows further stigmata of a degenerative kind in the form of polydactyly, having double thumbs, one of which has been amputated in infancy. The rickety condition is now quiescent.



Case No.10. Example of Cachectic Infantilism.

Sex.Male. Grade. Idiot. Age 21.

Family History.

Father,aged 42,health good,height normal.

Mother,aged 43,health good,height normal.

No history of mental defect or insanity in the family.Tuberculosis on father's side. No other relatives of small stature.

Siblings. Second child of four. Others healthy.

Birth normal. Wt.6 lb 11 oz. Never walked or talked. Teething normal. Defect observed at 1½ yrs. He was then said to have spastic paraplegia,squint, and microcephaly. Had fits since 18 months,mastoidectomy aged 2 yrs.,always frail and subject to respiratory illness every winter. Has the habits and mannerisms of a child of six months.

Physical Examination.

A tiny, emaciated person with a small head, thin body and slightly contracted legs. Genu valgum. Height 46 ins. Vertex-Symphysis 22 ins. Symphysis-Soles 24 ins. Span 48 ins. Circumference of head 18 ins. Weight $34\frac{1}{2}$ lb.

Fat. Very emaciated. Has a wizened look.

Posture. Cannot stand alone. Marked genu valgum and bilateral pes cavus. Slight mid-dorsal kyphosis.

Skin. Dry, wrinkled and mottled on face.

Teeth are large and widely spaced. Fairly regular with large central incisors. Eyes. Left internal strabismus. Fundi normal. Hair. Healthy on scalp.

There is no pubic or axillary hair. No thyroid palpable. No enlarged cervical glands.

Chest narrow. Expansion poor, resonance equal on each side. Medium crepitations scattered throughout.

Central Nervous System. Spastic quadriparesis.

Greatly increased deep reflexes. Bilateral plantar extensor reflexes. He shows "grasping and groping" reflexes like a baby.

Genitalia. Penis $1\frac{1}{2}$ ins. Small scrotum with both testes down, about the size of large peas.

Laboratory Findings.

B.P. 90/50. Urine normal. No phenyl pyruvic acid.

Blood and C.S.F. Wassermann reaction negative.

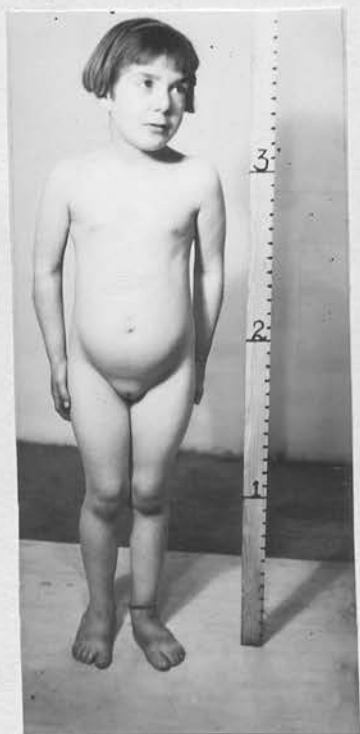
R.B.C's 3,300,000 c.m. Hb. 63% (Sahli) C.I. 0.9.

Radiologically, all epiphyses un-united. Bone age 11-12 years. Histologically. Endocrines not definitely abnormal except immature testes.

Summary.

A case of cachectic infantilism without any definite endocrine dysplasia. The glands, examined histologically, did not show any gross abnormalities with the exception of the testes which were very immature. There seems to have been an all round hypoplasia of the endocrines accompanying the general hypofunction of all body cells. The case shows some points in common with progeria but did not run the same course.

As a child, he was badly crippled from birth, the cause being genetic and showing itself as a spastic quadriplegia. The mentality reached only to the level of a six months old baby. Epilepsy began at two years after an operation for radical mastoidectomy. All through childhood he was continually ill with respiratory affections. All these factors combined to produce a tiny, infantile dwarf with no recognizable endocrine dysplasia. He died, aged 21, of pneumonia and the lungs showed chronic emphysematous changes. All the organs were undersized but not grossly abnormal. The head was not truly microcephalic, merely being reduced in all diameters in proportion and therefore nanocephalic. (See discussion on microcephaly.)



Case No.11. Example of Coeliac Dwarfing.

Age 10. Grade. Imbecile. Sex.Female.

Family History.

Father,aged 56,health good. Ht.66 ins. Wt.145 lb.

Mother,aged 49,health good. Ht.63 ins. Wt.159 lb.

Grandmother died of alcoholism and dropsy.

Siblings. She is the eighth child of thirteen.

Three brothers and ten sisters. All normal except

No.3,a male,who died in infancy of tuberculous meningitis. No other relatives of small stature.

Personal History.

Birth weight 8 lb. No forceps. Post partum haemorrhage. Walked at 5 yrs. Talked at 5 yrs. Teething was backward. Suffered from coeliac disease from three weeks of age. Nearly always in hospital.

Had diphtheria, measles, tonsils and adenoids and chicken-pox.

Habits. An odd, "old fashioned" little girl who is rather spoilt because of her long hospitalisation. Appears more intelligent than she really is.

Physical Examination.

Small, with a protruding abdomen and rather muddy complexion.

Height 46 ins. Standard height 53 ins.

Weight 50 lb. Standard weight 63.6 lb.

Vertex-Symphysis 24 ins. Symphysis-Soles 22 ins.

Span 44 ins. Fat. Moderately adipose with a protruding abdomen. Posture. Slight lordosis. Walks well. Skin. Smooth and supple but muddy.

Teeth. First dentition still present, worn down but fairly regular. Eyes. Slight right internal strabismus. Conjunctivae healthy. Fundi normal.

Ears. Slight discharge from right, due to eczema of meatus. No perforation. Thyroid impalpable and no enlarged cervical glands.

Chest. Some protrusion of sternum. No costo-chondral beading. Lungs healthy.

Abdomen. Protruberant and soft. Liver not enlarged but spleen is just palpable.

Genitalia. Immature. No secondary hirsuties.

No other abnormal clinical findings.

Laboratory Findings.

B.P. 115/75. Urine. No abnormal constituents.

Blood and C.S.F. Wassermann reaction negative.

Blood Sugar Curve shows slightly increased tolerance.

Laboratory Findings (contd.)

Blood cholesterol 144 mgms %.

Serum calcium 9.9 mgms%. Inorganic Phosphate 4.0mgm%.

Faecal fat. Total 15.08 gms %. Split - 69.8 %.

Unsplit 30.3 %.

Blood.R.B.C's 4,080,000. Hb.70%. C.I. 0.8.

W.B.C's. 16,700. Differential count. Polymorpho-nuclears 78 %. Lymphocytes 22 %.

Bone Age. Under nine years. Pisiform has not appeared. Widely separated carpals. Lower epiphysis of ulna just appearing.

Summary.

An odd, precocious, little girl who appears more intelligent than she really is. Intelligence quotient is 44 %. There is a history of tuberculosis and alcoholism in her family. She has spent nearly all her life in hospitals, either for physical or mental treatment. The coeliac disease is now cured but the sequelae remain in her dwarfed stature, pot-belly, and delayed ossification and dentition.

The faecal analysis is normal now.

Blood counts show a slight anaemia and a rather high white count. The spleen is slightly enlarged as it may be in rickets, and there is no reason at present to associate this with the blood findings. The cholesterol and blood sugar curves show a tendency to hypothyroidism. She remains well with attention to diet and calcium and vitamin D added, especially in winter.



Case No.12. Example of Hypopituitarism.

Sex.Male. Grade.Idiot. Age 21.

Family History.

Father,alive and well. Normal height.

Mother has been lost sight of. Not unduly small.

Maternal grandmother was epileptic. No insanity or mental defect in family.

Siblings. Patient is second child of two. Elder brother is alive and well,and of normal height.

Personal History.

Birth was instrumental. He was late in walking and never talked. Teething normal. Still incontinent.

Epileptic from infancy. Fell from pram aged six months.

Physical Examination.

General Appearance. A restless, hyperkinetic boy quite withdrawn and inco-operative. Small, slim and of infantile type.

Height 56 ins. Vertex-Symphysis $27\frac{1}{2}$ ins. Symphysis-Soles $28\frac{1}{2}$ ins. Span $55\frac{1}{2}$ ins. Weight 67 lb.

Fat. Little anywhere. Posture. Can stand erect but it is difficult to get him to do so. Skin is smooth and fine but has many bruises. Teeth. Upper teeth have been removed. Lower teeth are quite good and well spaced. Hair glossy and fine. He has scarcely any pubic or axillary hair. Eyes healthy. Fundi could not be examined. No thyroid palpable. No enlarged cervical glands.

Chest is quite well formed. Spine straight. Lungs healthy. No gross abnormalities in the other systems. Genitalia are very small and immature. Both testes are descended but are small and soft.

Laboratory Findings.

B.P. 90/65. Urine. No abnormal constituents.

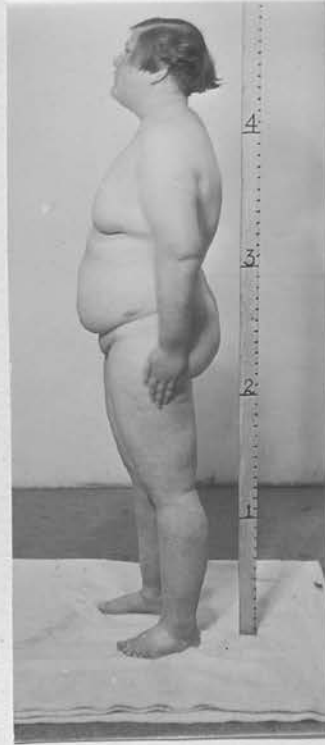
Wassermann reaction negative. Blood sugar curve is normal. R.B.C's 4,200,000 per c.m. Hb. 70 %.

C.I. 0.83. Radiological Examination shows a small hooded sella turcica with approximated clinoid processes. The bone age is under sixteen years. Carpals unmassed. Lower epiphyses of ulna and radius not fused. Upper epiphyses of radius and ulna wide open.

Summary.

This boy shows the usual features of what is known as the Lorain-Levi type of pituitary infantilism. (1871) He is slim and undersized with a smooth, white skin. The facial features are well formed and, though he is an idiot, look normal. The proportions are good, the lower being one inch more than the upper. The hands are well shaped and slim. Secondary sex characteristics are grossly retarded although both testes are descended. Those teeth which he has left are regular and fairly well calcified. There are no signs of hypothyroidism.

Radiological examination shows the sella to be small and enclosed by the clinoids. The osseous development shows the same retardation as the stature and sex organs and the bone age is about fifteen years. All this is due to hypofunction of the anterior pituitary with consequent lack of growth and gonadotropic hormones.



Case No.13. Example of Fröhlich's Syndrome.

Sex.Female. Age 19. Grade.Imbecile.

Family History.

Father,aged 45,health good. Ht.65 ins. Wt.140 lb.

Mother,aged 45,health good. Ht.66 ins. Wt.126 lb.

History of mental deficiency on father's side.

Epilepsy and tuberculosis on mother's side.

Siblings. The third child of eleven. 6 brothers and 4 sisters. Two brothers died in infancy and another is a mental defective. Others normal.

The grandparents were only about five feet high.

Personal History.

Birth normal. Wt. $7\frac{1}{2}$ lb. Walked at two years.

Talked at two and a half years. Teething normal.

Illnesses. Tonsils and adenoids removed, aged 7.
 Treated for thyroid obesity, aged 10-11.
 Habits. Marked behaviour disorder. Certified as defective because she was uncontrollable at home. Spiteful to children and animals and is much given to putting foreign bodies in her ears.

Physical Examination.

General Appearance. A very fat girl with pectoral and girdle obesity of pituitary type. Marked genu valgum and acrocyanosis of extremities.
 Height. 57 ins. Vertex-Symphysis $29\frac{1}{2}$ ins. Symphysis-Soles $27\frac{1}{2}$ ins. Span 58 ins. Chest and Hips both 41 ins. Waist 37 ins.
 Fat mainly around the chest, hips and proximal halves of limbs. Posture erect. Genu valgum.
 Skin. Soft, pliable, bruises easily, acrocyanotic.
 Hair. Fine and silky. Very slight amount of pubic and axillary hair. Eyes healthy. Fundi normal.
 Teeth are slightly crowded but regular and well calcified. Neck very fat. Double chin. No thyroid palpable. Breasts show fat deposition but little glandular tissue. Genitalia are immature and she has not yet menstruated at age of nineteen.
 No other clinical findings of note.

Laboratory Findings.

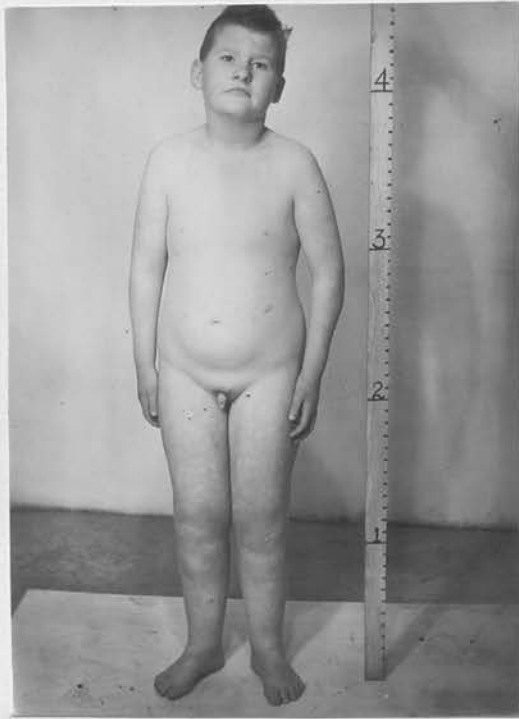
B.P. 130/85. Urine. No abnormal constituents. Wassermann reaction negative. Blood chlorides 458 mgms NaCl %. Sugar Tolerance Curve is of "lag" type. No polyuria. Serum Calcium 9.2 mgms %. Inorganic Phosphate 4.2 mgms %.
 X-ray - see discussion.

Summary.

A case of pituitary obesity of the Babinski-Fröhlich type. She has a bad family history from the mental deficiency viewpoint. She herself is an imbecile who shows a marked behaviour disorder. She is incredibly naughty and quite unamenable to discipline of any kind.

She shows the salient points of this type of pituitary deficiency. Werner (1942) blames the posterior pituitary while Grollman (1941) stresses the hypothalamic aspect. The cause, in her case, may be due to a cranio-pharyngioma. There is a calcified area in the sella which is to be further investigated. The secondary sex characters are still almost undeveloped and she has never menstruated. She shows the easy bruisability met with in these cases. (see picture). Height is not much interfered with in this type although they seem never to be tall. Osseous development is delayed by about three years in her case. Bone age 16 years.

The blood chemistry figures are not abnormal and she has no polyuria. Her hands are typically plump and tapering. She has shown some diurnal somnolence but there are no signs of eye disturbances in the way of optic atrophy or bitemporal hemianopia.



Case No.14. Example of Hypopituitarism. Sex Male.
Age 12. Grade Imbecile.

Family History.

Father, aged 35. Health good. Ht. $72\frac{1}{2}$ ins. Wt 196 lb.

Mother, aged 34. Health good. Ht. 68 ins. Wt. 147 lb.

Siblings. The patient was the first born. Two younger sisters are normal and very intelligent.

No relatives of very small stature.

Personal History.

Birth normal. Weight $7\frac{1}{2}$ lb. Walked at three years.

Teethed early, talked at four years.

Illnesses. Measles at 5 years. Was treated for hypopituitarism during childhood. Suspected fracture of the skull at 2 years.

Habits. A very naughty little boy. Mischievous and spiteful to other children. Had to be transferred to the Male Side of the hospital at the age of ten because he was unmanageable by female staff.

Physical Examination.

General Appearance. A fat boy with a fresh complexion and acrocyanosis of extremities. Very small genitalia.

Weight 123 lb. Height 54 ins. Symphysis-Vertex 28 ins. Symphysis-Soles 26 ins. Span 53 ins.

Fat. Very adipose especially around the hips.

Posture erect. Walks and runs normally. Slight genu valgum.

Skin is fine and smooth. Bruises easily. Marked acrocyanosis, especially of lower limbs.

Hair soft and silky. No axillary hair. Pubic hair just beginning to appear.

Mouth. Dentition is slightly retarded. The second molars have not yet appeared and the canines and second premolars are not long erupted. The teeth are fairly well calcified and regular.

Eyes healthy and fundi normal. Neck is fat and no thyroid is palpable.

Breasts. Some deposition of fat. No real evidence of femininity.

Respiratory System shows no abnormalities.

Cardio-Vascular System. B.P. 90/50. Otherwise normal.

Alimentary System. Abdomen obese. No enlargements.

C.N.S. Nil abnormal except all reflexes sluggish.

Urine. No albumin, sugar or phenyl pyruvic acid.

Genitalia. The appearance suggests that the patient

narrowly escaped being a male pseudo-hermaphrodite. The small penis, ($1\frac{1}{4}$ ins), and tiny scrotum, are surrounded by an oval pad of fat which, in its upper part forms a mons veneris. The right testis is palpable outside the abdominal ring and is about the size of a hazel nut, and the other is undescended.

Laboratory Findings.

Wassermann Reaction negative. Serum calcium 9.9mgm%. Inorganic Phosphate 4.8 mgms%. Sugar Tolerance curve is of mild diabetic type. Ossification is not significantly delayed. X-ray of the sella shows it to be short antero-posteriorly and to be almost bridged by the anterior clinoid processes which run down and back to the posterior ones.

Summary.

This boy shows obesity and sexual retardation of hypopituitary type, approximating to the extent of the preceding case. Since the examination set out here, however, he has kept on losing weight, has grown more pubic hair and has a distinctly palpable right testis. The genitalia are slightly bigger. He is therefore beginning to overcome his former gross degree of hypopituitarism without treatment and is not a true Babinski-Fröhlich type although his condition earlier was indistinguishable from this.

His hypopituitarism has led to some retardation of stature, delayed dentition, and ? delayed ossification. The sella is abnormal in shape and

is very short antero-posteriorly and deep. There is no intra-sellar calcification or intra-cranial pressure. The fundi are normal and the eye-fields full. He had never had any glycosuria on routine urine tests but the mild diabetic sugar tolerance curve suggests hypothalamic interference. Owing to his behaviour disorder it was not possible to perform water tolerance tests. There is no evident polyuria. It is proposed, should the sexual improvement and loss of weight not continue, to aid him with a course of pregnancy urine or serum extract.



Case No 15. Example of Untreated Cretinism.

Sex, Female. Age 37. Grade, Idiot.

Family History.

No details about the parents available.

The patient was the second of a family of eight.

The others are said to be normal.

Two cousins are in mental hospitals. One of these is in this hospital. She shows no endocrine dysplasia but is an idiot with a primitive kind of schizophrenic psychosis. The patient lived at home and never appears to have had any sustained thyroid replacement therapy.

Illnesses. Always frail and semi_invalid, spending a great part of her life in bed and contracting chest trouble every winter.

Physical Examination.

General Appearance. A tiny, twisted, little woman with cretinoid features, kypho-scoliosis, pot-belly and pronounced genu valgum.

Height 40 ins. Weight 66 lb. Symphysis-Vertex 22 ins. Symphysis-Soles 18 ins. Span 37 ins.

Fat. Protuberant abdomen but only moderately covered with fat. Well marked neck and shoulder pads.

Posture. Can stand with slight assistance. There is marked twisting of the spine and moderate flexion at the hip joints.

Skin thick and rather harsh. **Hair** dry and lustreless but thick. **Mouth.** Teeth poorly calcified, irregular, crowded and carious.

Eyes. Some conjunctivitis. Vision could not be tested owing to inco-operation. **Neck.** No thyroid or cervical glands palpable.

Chest. Movements very poor. Ribs splayed by the protuberant abdomen. Percussion fairly resonant and equal. Prolongation of expiration and scattered medium and coarse crepitations through both lung fields.

Cardio-vascular System. Pulse 60 per min. Regular but feeble. B.P. 102/74. Poor peripheral circulation. Heart sounds faint, regular and closed in all areas. **Alimentary System.** Tongue rough and dry. Abdomen protuberant but no enlargements felt. Small umbilical hernia.

C.N.S. The patient is dumb. Pupils are equal, active and circular. All reflexes present, equal but sluggish. Can walk with assistance.

Urine. No albumin, sugar or phenyl pyruvic acid.

Genitalia. Atrophic. Very sparse pubic hair.

Axillary hair present in small quantity. No other hirsuties. Complete amenorrhoea.

Breasts. Flat and flabby. Small, immature nipples.

Laboratory Findings. Wassermann Reaction negative.

R.B.C's. 3,980,000. Hb (Sahli) 71%. C.I. 0.9.

W.B.C's. 8,200. Polymorphs 68%. Lymphocytes 28%.

Hyalines 4%.

Post Mortem Examination.

1. Thickened dura mater adherent to pia.
2. No thyroid tissue at all discovered.
3. Confluent broncho-pneumonia.
4. Marked atheroma of aorta and coronary vessels.
5. Very small spleen. (50 grammes)
6. Lobulated kidneys. Left kidney cystic with dilated pelvis.
7. Uterus and ovaries very small. Cyst of left ovary.
8. Sub-periosteal haematoma of left knee joint.

The other endocrines were normal macroscopically but were not sectioned.

Summary.

A cretin of classical type who had never had any consistent substitution therapy. She survived to the age of 37 in sheltered circumstances. She showed a great sensitivity to thyroid treatment and the dose had to be constantly regulated and often decreased.



Case No.16. Example of Partially Treated Cretinism.

Sex,Female. Grade.Imbecile. Age 15.

Family History.

Father.No facts known. Illegitimate.

Mother.Age 38.Health good. Ht.62 ins. Wt. 151 lb.

Siblings. The first child of four. The others are step brothers and sisters. All normal.

No relatives of small stature.

Personal History.

Birth difficult. Large head. No forceps. Wt 8 lb.

Walked at 2 years,talked at three years,teething
? normal.

Illnesses. Whooping cough,measles and chicken-pox.

Habits. A shy,pleasant little girl who can do
simple sewing. Immature.Plays with younger children.

Physical Examination.

General Appearance. She looks about eight years of age. The stigmata of cretinism are not plainly evident.

Height 46 ins. Standard height for age 62.5 ins.

Weight 62½ lb. Standard weight 107 lb.

Symphysis-Vertex 24½ ins. Symphysis-Soles 21½ ins.

Span 46 ins.

Fat. Well covered. No girdle distribution.

Posture erect. Walks and runs normally.

Skin smooth. Teeth regular and quite well calcified. Hair glossy and healthy. Eyes healthy. No squint. Fundi normal. Neck. No thyroid or cervical glands palpable. Breasts immature.

Respiratory System. Not abnormal.

Cardio-Vascular System. Pulse 90. Regular in time and force. B.P. 118/70. Heart sounds pure and closed.

Alimentary System. Tongue smooth and clean.

Abdomen rather fat. Nil palpable. No herniae.

Nervous System. Pupils equal, circular and active.

All reflexes present, equal and brisk.

Urine. No albumin or sugar.

Genitalia quite immature. No trace of pubic or axillary hair. Menses not begun.

The hands and feet are relatively very large as compared with the rest of the physique.

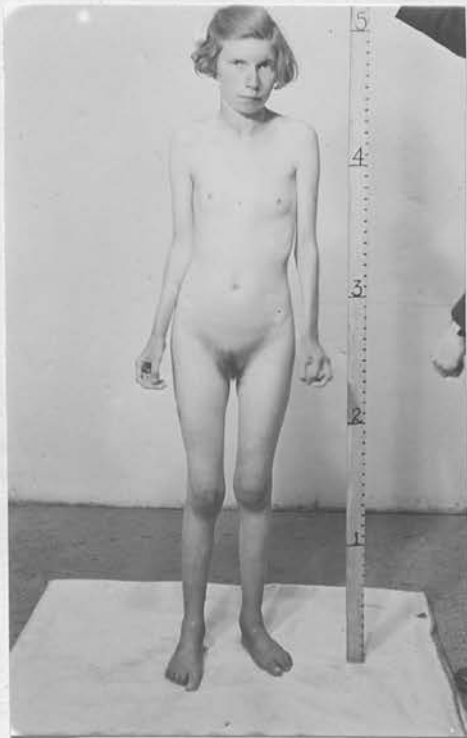
Laboratory Findings. Wassermann negative. Blood cholesterol 156 mgms%. Delayed ossification.

Carpal bones widely separated and os pisiform has not yet appeared. Lower epiphysis of ulna has just appeared. Bone age about 8 years. Sella turcica normal.

Summary.

An example of cretinism in a girl of fifteen who has had inadequate substitution therapy. This has resulted in her appearance being improved to an extent which makes it difficult to appreciate her cretinoid features at first glance. Together with the hypothyroidism there is no doubt an accompanying underfunction of the pituitary and other endocrines. Withdrawal of thyroid over a period of two months resulted in a reduction of pulse rate to 70 per minute and a raising of the blood cholesterol to 190 mgms%.

She has a rounded face, flattened nose, and relatively large and broad hands and feet. Substitution therapy has not been adequate enough to bring about increase in stature or to prevent delay in the union of epiphyses and appearance of centres of ossification. Mentally she appears brighter than she really is and her intelligence has suffered considerable stunting, her intelligence quotient being only 40% of normal. Puberty is greatly delayed and there are no secondary sex characters as yet. This is more likely to be due to an accompanying hypopituitarism.



Case No.17. Example of Hypogonadism. Sex.Female.
Age 19. Grade.Idiot with primitive type of schizo-
phrenia.

Family History.

Father.Age 58.Healthy.Height 69 ins.Weight 142 lb.

Mother.Died aged 54.Cancer. Ht.66 ins. Wt.unknown.

No relatives of small stature.

Siblings. 1.Twins,male and female,normal.

2. Female,normal.

3. Twins.F.normal. M.died 3 months.

4. Patient.

Personal History.

Normal birth. Weight 7 lb. Walked at 5 years.

Talked a little at 9 years. Teething normal.

Illnesses.Measles,heart attacks,dysentery,aet 16.

Habits. A schizoid girl who rarely speaks. She exhibits catatonic features such as *flexibilitas cerea* and does nothing to help herself. She is doubly incontinent and cannot be trained to be clean in habits.

Physical Examination.

General appearance. A thin, angular girl who stands or sits apathetically where she is put. She has well marked acrocyanosis of hands and feet.

The breasts are undeveloped and the pubic hair is sparse.

Weight 92 lb. Standard weight 122 lb.

Height 63 ins. Symphysis-Vertex $30\frac{1}{2}$ ins. Symphysis-Soles $32\frac{1}{2}$ ins. Span 64 ins.

She is very lean with thin limbs. Posture is poor and she slouches. Skin is smooth, cold and shows cyanosis and chilblains. Hair is lustreless.

Mouth. Strong healthy teeth. Large central incisors and small lateral incisors. Eyes healthy and fundi normal. Neck thin. No thyroid or cervical glands palpable. Breasts quite immature with flat nipples.

Respiratory System. Chest moves badly. Resonance is equal bilaterally and there are no crepitations.

Cardio-Vascular System. Pulse 96 per min. Regular.

B.P. 125/85. The heart is not enlarged. There is a thrill over the whole praecordium. A systolic murmur prolonged into diastole is heard best over the mid-sternum. Blood count. R.B.C's 4,800,000 per c.m. Hb. 88%. (Sahli) C.I. 0.9.

Alimentary System. Tongue is clean and smooth.

Abdomen lax. Liver and spleen not palpable.

Central Nervous System. No abnormalities.

Urine. No albumin or sugar.

Genitalia. Hypoplastic. Thin pubic hair. Axillary hair just appearing. Primary amenorrhoea.

Laboratory findings. Wassermann Reaction negative.

Normal Sugar Tolerance Curve. X-ray of hands. The epiphyses are not fused. Phalanges long and tapering.

Phalangeal epiphyses and lower epiphyses of radius and ulna still open. Bone age 15-16 years.

Sella is normal in outline.

Summary.

A girl of nineteen years who shows a skeletal make-up indicative of gonadal insufficiency. X-ray of the sella is normal. She has not yet menstruated. Her limbs are slim, the symphysis-soles measurement exceeds the upper half of the body, and the span is greater than the height. The teeth show typical hypogonadal features, the central incisors being large and the lateral incisors small. The genitalia are immature, secondary sex characters are poorly developed and there is some retardation of epiphyseal closure.

She has a congenital heart lesion in the form of a patent interventricular septum. There is no cyanosis of lips or clubbing of fingers, only a marked acrocyanosis of hands and feet which is seen regularly in schizoid persons and defectives.

Case No.18. Example of Mongolism with Diabetes.
Sex.Female. Age at Death.18 yrs. Grade.Imbecile.

Family History.

Health of parents good. They are of normal size.
There is no history of diabetes,insanity,mental deficiency,epilepsy,tuberculosis or alcoholism to be found in the family.

Siblings. Brother,aet 23. Healthy. Scholarship.

Sister,aet 20. Healthy.

Brother d. in infancy after circumcision.

Patient.

Personal History.

Birth was normal. Walked at 18 months. Teething normal. Backward in talking. Deficiency was first noted at five years! Habits always faulty.

Illnesses. Measles,scarlet fever,bronchitis, otorrhoea and diabetes mellitus. Attended hospital weekly as an out-patient until admission to mental hospital,aged fourteen.

Habits. She was so naughty at home that her parents were unable to control her and keep her from stealing food. It was thought that her diet could be more carefully controlled in a colony. She was always overactive,noisy and disobedient.

Physical Examination.

General Appearance. A thin little girl with permanent blepharitis and typical Mongolian features.

Weight 74 lb. Height 54 ins.

Standard weight 117 lb. Standard height 64 ins.

Fat. Very little anywhere.

Posture. Erect with a slight lordosis.

Skin dry and harsh. Hair dull and lifeless.

Mouth. Hypoplastic upper incisors. All teeth rather small and poorly calcified, with ridged enamel.

Eyes. Internal strabismus of left eye. No cataract.

Permanent blepharitis. Fundi, no definite changes.

Neck. Thin. No thyroid or cervical glands palpable.

Respiratory System. Bilateral bronchitic signs.

Cardiovascular. Pulse 88 per min. Regular.

B.P. 130/90. Heart sounds forcible but pure and closed.

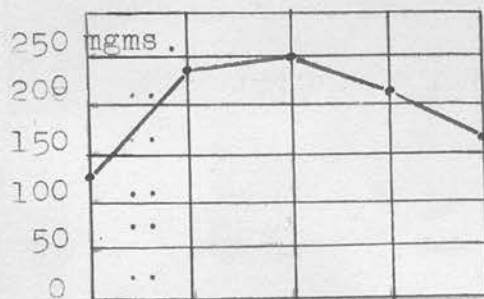
Abdomen. Lax. No enlargement of liver or spleen.

C.N.S. Strabismus as above. No other lesion detected except the generalised hypotonia usual in Mongolian Imbeciles.

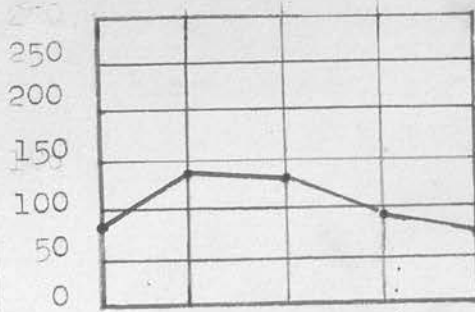
Urine. Often contained sugar and acetone when she stole extra food or was ill.

Genitalia. Pubic and axillary hair sparse. Genitalia otherwise normal, but menses, which began at 16 years, were always irregular and scanty.

Laboratory Findings.



On admission.



On 1260 calories
and 15 units T.I.D.

Glucose Tolerance Curves after 50 grams Glucose.

Progress.

Insulin dosage varied between 7 and 18 units T.I.D. during the four years which she spent in hospital. She had several hypoglycaemic attacks and, during an attack of pneumonia, she became comatose and had to receive intravenous glucose. A year after this when she was really very well on 18, 18, and 15 units daily she contracted a second attack of pneumonia and in spite of urgent measures, died in three days. Post mortem was refused.

Summary.

A case of Mongolian Imbecility with diabetes. This is rather rare as Lawrence (1942) points out. The case is included here to show that a Mongol even with a severe accompanying disorder such as diabetes mellitus, may not show reduction of stature (as compared with other Mongols) and may reach puberty without much delay. It would be expected that an endocrine disturbance such as diabetes would interfere markedly in such a person as this disease often leads to irregularity and amenorrhoea in otherwise normal women.



Case No.19. Example of Mongolism with Congenital Heart Disease. Sex.Male. Age 14. Grade. Idiot.

Family History.

Father,aged 68.Health good.Ht.69 ins. Wt.196 lb.

Mother,aged 58.Health poor.Liver and kidney trouble. Height 66 ins. Weight 154 lb.

Siblings. Nil. He is an only child.

No other relatives of small stature.

Personal History.

Birth.Long labour with vomiting. Weight 4 lb.

Mother aged 44 years at time of birth.

Walked at 5 yrs.Never talked.No teeth until he was $2\frac{1}{2}$ yrs.

Illnesses. Bronchitis every spring and autumn.

Habits.Quiet and inoffensive but restless.

He is unclean in habits.

Physical Examination.

General Appearance. A restless, vacant looking little boy with typical Mongolian features.

He has double squint and acrocyanosis.

Height. 49 ins. Standard height for age 60.7 ins.

Weight. 66 lb. Standard weight for age 92.9 lb.

Symphysis-Vertex 26 ins. Symphysis-Soles 23 ins.

Span 46 ins.

Fat. Well covered. Posture erect. Skin smooth and fine. Acrocyanosis of extremities. Hair strong and wiry. No axillary hair. No pubic hair.

Mouth. Teeth widely spaced. All four lower incisors extracted before admission. Congenital absence of the upper lateral incisors. Delayed eruption of second upper molars.

Eyes. Slight blepharitis. Double internal strabismus. Fundi normal. Neck. No thyroid palpable.

Respiratory System. Protruding sternum with enlargement of the costo-chondral junctions.

Both lung fields show signs of chronic bronchitis.

Cardiovascular. Pulse 90. per min. Regular in time and force. B.P. 108/70. The heart shows slight enlargement to the right and there is some cyanosis of lips and extremities. No finger clubbing. A harsh systolic murmur can be heard over the whole praecordium. There is a thrill palpable over the base. The condition appears to be one of patent interventricular septum with slight stenosis of the pulmonary artery.

Other Systems not abnormal. Urine clear.

Genitalia are minute. No secondary sex characters.

Summary.

This is a case of Mongolian Imbecility with accompanying congenital heart disease. The Wassermann reaction is negative. The patient shows most of the usual stigmata associated with the condition. He has a brachycephalic head, tilted palpebral fissures, blepharitis, squint and short buccal cavity with large fissured tongue. The eyes have moderately well marked epicanthic folds. The limbs are short in comparison with the body and are extremely lax and mobile in the joints. The hands and feet are short and stubby, the fifth finger is in-turned and does not reach to the second crease on the ring finger. The palms show the simian crease running straight across the palm.

All the above facts apply equally well to most of the Mongols in the group, but here we have in addition, a congenital heart lesion which has further retarded growth in an already stunted physique and led to a definite amount of sexual hypoplasia. Mongols of a like age are both taller and show secondary sex changes comparable with normal boys of that age.

Uncomplicated Mongolism does not appear to lead to sexual retardation, reduction of total height being the only "infantile" character.

Case No 20. Example of Sexual Hypoplasia accompanying Cerebro-macular Degeneration.

Sex.Female. Age at Death.19 yrs. Idiot.

Family History.

Father and mother are both healthy, of average height and weight, and not related.

Siblings. Elder brother, aged 22. Healthy.

Twin brother of patient, aged 19, healthy.

No history obtained from the parents of any other cases in the family. Follow-up shows the other children to be still healthy.

Personal History.

Birth was normal. Weight not known.

Teethed, walked, talked and was clean in habits at the normal time.

Illnesses. Chicken-pox, measles and mumps before the age of seven. Aged 7, she had an attack of scarlet fever. Two years after this her sight began to fail and she became very restless and anxious. When she was 11 she began to have fits.

Physical Examination.

General Appearance. Thin and small with contracted legs and right arm. Pale and restless. Blind.

Height 59 ins. Weight 96 lb. Symphysis-Vertex 29 ins. Symphysis-Soles 30 ins. Span 59 ins.

Emaciated. Posture. Cannot stand alone. Legs are contracted. Skin pale and moist.

Mouth. Irregular dentition, caries, spongy gums and hypoplastic incisors.

Physical Examination (contd)

Eyes. Vacant and staring. Fundi. Pale discs with indistinct edges. Arteries very shrunken.

Scattered areas of pigmentation and irregular mottling with choroidal atrophy.

Neck. No thyroid palpable.

Respiratory. Chest thin and narrow, no other gross abnormalities detected.

Cardiovascular System. Pulse 32, per min. Poor volume. Regular in time and force. B.P. 110/60.

Heart sounds faint, but pure and closed.

Alimentary System. Tongue furred. Abdomen thin. No enlargement of liver or spleen.

Urine. Contained albumin at times, with blood cells and pus, but no casts. Clear for six months before death.

Central Nervous System.

Pupils are fixed and do not respond to light.

Fundi - see before. There is advanced involvement of the pyramidal tracts with spasticity and flexor contracture of both legs and the right arm. All jerks are now difficult to elicit owing to these contractures. Abdominal reflexes are absent and both plantar reflexes extensor.

She has difficulty in swallowing and has to be fed on slops.

Genitalia are hypoplastic, pubic and axillary hair very sparse. Menstruated at 15, but was very irregular and scanty and has now had amenorrhoea for the past year. Breasts small and immature.

Wassermann Reaction negative.

Summary.

Post mortem examination of the brain in frozen sections in this case showed signs typical of cerebro-macular degeneration. (Waren Tay-Sachs' Disease). There was degeneration with swelling of the cell protoplasm, chromatolysis, destruction of cell fibrils, disappearance of cell nuclei and lipoid accumulation. Atrophy of the cerebellum, which is often found, was well seen.

This is the juvenile type where onset was delayed until the age of eight, and appeared to follow an attack of scarlet fever. There is no evidence of any other member of the family being affected and this is especially interesting as the patient was one of twins. Both the retinal changes and the histological findings bear out the diagnosis which was made some years before death by the late Dr. Kinnier Wilson.

The brain trauma in the pre-pubertal period led to some stunting of growth (59 ins.) and to a condition of sexual retardation and immaturity shown by the failure of development of the breasts, poorly defined secondary sex characters, rather late onset of menstruation and early cessation at eighteen.



Case No.21. Example of Dwarfing and Infantilism following on Epidemic Encephalitis.

Sex.Female. Age 24. Grade.Idiot.

Family History.

Father,aged 50,health good. Ht 70 ins.Wt.147 lb.

Mother,aged 50,health good. Ht.68 ins.Wt.142 lb.

No history of insanity or defect in family.

Siblings. Patient is the fourth child of five.

The others are all healthy and normal.

There are no relatives of small stature.

Personal History.

Birth normal.Wt 7 lb. Walked at $1\frac{1}{2}$ yrs. Talked at $1\frac{1}{2}$ yrs. Teething normal.

Illnesses. "Sleeping sickness" at five years.

Habits. She is now an incontinent,dribbling idiot.

Physical Examination.

General Appearance. A thin, white, crippled girl.

Small stature and contracture of right leg.

Height 51 ins. Weight 61 lb. Symphysis-Vertex

24 ins. Symphysis-Soles. 27 ins. Span 52 ins.

She is thin almost to the point of emaciation.

Hair is dry and lifeless. Teeth are surprisingly good. The incisors have been removed.

Eyes. Pupils are unequal, right larger than left.

Reaction to light sluggish. Accomodation cannot be tested. No opacities and the conjunctivae are healthy. Skin is pale and rather greasy.

Neck. No thyroid or cervical glands palpable.

Breasts. Small, with flattened nipples. No glandular tissue. The small bulk being made up of soft fat.

Respiratory System. There is some kypho-scoliosis and the chest is small and cramped especially on the left side. Lung fields clear.

Cardiovascular System. Pulse 80. Regular in time and force. B.P. 125/85. Heart slightly enlarged to the left, and there is a mitral systolic murmur maximal at the apex.

Blood Count. R.B.C's. 3,200,000 per c.m. Hb. 62%.

C.I. 1.0.

Abdomen is held rigid. Nothing palpable.

Urine. No abnormal constituents.

C.N.S. Pupils as above. She has generalised spasticity of all limbs but the right side is most affected and the right leg is contracted with talipes equinus of the foot. All reflexes are very exaggerated except those of the contracted

limbs. Abdominal reflexes are absent. Plantar reflex is extensor on the right side. She has slight tremor of the hands and sometimes shows pill-rolling movements. Her face is usually solemn and when she laughs the grin appears and disappears slowly.

Genitalia. There is only sparse pubic hair. The external genitalia are immature and there is no mons veneris. Axillary hair is practically absent. She has never menstruated.

Wassermann Reaction negative., in blood and C.S.F.

Summary.

A normal child until the age of five. Then had an attack of epidemic encephalitis. This led to a gross behaviour disorder in the next three years. At eight she was deemed feeble-minded and was then very violent and destructive. Her right-sided hemiplegia did not exhibit itself until she was ten or eleven years of age. At this time she had a protracted illness with an irregular temperature. She has the appearance of a post-encephalitic and the history of the illness is clear, but she has pyramidal involvement not seen in a pure case of Parkinsonism.

Her stature, (51 ins) and sexual immaturity seem to be directly due to the effects of this crippling illness.



Case No.22. Example of Retrograde Infantilism.

Sex, Male. Mental Grade. Imbecile. Age 34.

No Family History is available in this case.

Personal History.

Deficiency observed from early infancy and presumed to be of congenital origin.

Said to have a congenital heart lesion in 1926.

There is no evidence of this now.

Alopecia developed in 1930 when the patient was 22, and at that time he was noted as having only sparse pubic and axillary hair.

During 1930-32 he had fairly frequent fainting attacks but these have been absent for the past four years.

In 1937, when he was 29 years old, he was first noted to have small testicles. No previous illnesses.

Physical Examination.

General Appearance. He has a Mongolian look but does not show the typical characteristics of Mongolian imbecility. He is completely bald and has no lower lashes and extremely sparse eyebrows especially in the outer third. His body length is greater than leg length, even to the naked eye. Height 60 ins. Weight 107 lb. Span 60 ins. Symphysis-Vertex 32 ins. Symphysis-Soles 28 ins. Fat, normal. Posture erect. Skin, fairly soft and pliable. Hair. Complete alopecia of scalp. Only has to be shaved once weekly and growth is weak. Eyebrows very sparse and almost absent in outer third. No upper eyelashes. Lower lashes short and stubby. Axillary hair thin, but long and straggling. Pubic hair. Fair amount with female distribution. Mouth. Irregular, carious teeth with high V palate. Eyes. No blepharitis. Fundi normal. Neck, long and sturdy. No thyroid or cervical glands palpable. Respiratory System. No abnormalities noted. Cardiovascular System. Pulse 48 per. min. Regular in time and force. Blood Pressure 110/70. Heart sounds pure and closed in all areas. Blood Count. R.B.C's. 4,850,000. Hb. 90%. C.I. 0.9 Abdomen rather full. Lax musculature. Nil palpable. C.N.S. Pupils react to light and accommodation and are equal and circular. All reflexes sluggish and equal. Urine. No abnormal constituents. Genitalia. The penis is of normal size and the scrotum is long and pendulous. The testes are

Physical Examination (contd.).

extremely small and soft, like large peas.

Erections are infrequent and masturbation, which was at one time practised, has been discontinued.

Rectally, the prostate gland is very small and soft in consistency.

Laboratory Findings.

Wassermann Reaction negative.

Blood Sugar Tolerance Curve. Normal.

Blood Cholesterol 154 mgms %.

B.M.R. (Reed's Formula) -14 %.

Epiphyses are all fused.

Goetsch Test, negative. (Werner, A. 1942. p 851.)

Urinary Chloride Excretion. (Cutler, Power and Wilder 1938.) 50.0 mgms %. Normal is 54 mgms %.

In hypoadrenia values are around 293 mgms %.

Summary.

This patient exhibits alopecia of the scalp, outer eyebrows and upper lids. Beard growth is greatly reduced. The axillary hair is sparse and the pubic hair female in distribution. The baldness developed at the age of 22. Since then he has had decreasing libido and the testes are now very small and soft and the prostate is barely palpable and of soft consistency. He has a history, after the baldness came on, of frequent fainting attacks for two years. These are now absent. The symptoms may be caused by hypothyroidism but the B.M.R. (Reed) is not unduly low. (It was not possible to do an accurate B.M.R.

owing to war conditions.) The blood sugar curve is not depressed and the blood cholesterol not unduly high. The pulse is, however, only 48 per minute, and the blood pressure low. Neuro-circulatory disturbances (the fainting attacks) are found in hypogonadism, and hypoadrenia may give a like picture. Werner (1942) quotes hypoadrenia as a cause of alopecia. In this case evidence of hypoadrenia was lacking by the chloride excretion test.

The case is shown as one in which there is regressive infantilism coming on after sexual maturity. The cause seems most likely to be hypothyroidic but whether this is the primary condition, or not, is not known.



Case No.23. Example of Dystrophia Myotonica with Regressive Sex Changes. Sex.Male. Age 33.

No family History is available in this case.

Physical Examination.

General Appearance. A dull,vacant looking man who shows frontal baldness,facial weakness with open hanging mouth,and muscular wasting.

Height 60 ins. Weight 94 lb. Span 59 ins.

Symphysis-Vertex 30 ins. Symphysis-Soles 30 ins.

Fat. Very lean,almost to emaciation.

Posture poor. Drooping shoulders. Some degree of genu valgum,and both feet deviate to the right.

Skin sallow and greasy. Hair. Frontal baldness with dry,scanty remaining hair. Pubic,axillary and chest hair normal.

Physical Examination (contd).

Eyes. There are signs of early cataract in the right eye. Fundi are not easily seen, but appear to be normal.

Neck is very thin. No thyroid palpable. The sterno-mastoid muscles are wasted.

Respiratory System. Chest moves badly. There is some flattening of the left apex. He has had attacks of bronchitis and was thought at one time to have pulmonary tuberculosis, but at present the condition is not found clinically or by radiography.

Cardiovascular System. Pulse 80 per min. Regular. B.P. 120/80. Heart sounds are pure and closed in all areas.

Alimentary. The teeth are very foul, irregular and covered with tartar. Abdomen is lax and no enlargements are felt on palpation.

The urine contains no abnormal constituents.

Central Nervous System.

The abnormalities present are due to the myopathy.

These are:-

(1) Slow relaxation of muscle after stimulus. E.g. Handshake or tapping tongue with hammer.

(2) Weakness and wasting of the muscles of the face with sad "myopathic" expression, drooping nether lip and open mouth.

(3) Wasting and weakness of sterno-mastoids.

(4) Flexor weakness of arms, wasting and weakness of the vasti of thighs and flat feet with deviation to the right, presumably due to weakness

of the dorsiflexors and allied muscles. There is no fibrillation to be seen.

(5) The voice is nasal and monotonous.

(6) Sensibility to pain, touch, heat and cold is unaffected.

(7) Pupils are equal, central, circular and active, reacting to light and accommodation.

Deep reflexes are all diminished but equal on both sides. Plantar reflexes are bilaterally flexor.

Genitalia. There is a good growth of pubic hair and the penis is of normal size but both testes are small and soft, about the size of beans.

Erections are not now seen and seminal emission absent.

Laboratory Findings.

Wassermann Reaction negative.

Serum calcium 9.5 mgms %. Inorganic Phosphate 2.45 mgms %.

Summary.

A typical case of Dystrophia Myotonica showing the cardinal signs of the disease such as myotonia, muscular wasting, premature baldness, cataract, bodily wasting and loss of sexual power with testicular atrophy.



Case No.24. Example of Microcephaly. Sex.Male.
Age 21. Grade.Idiot.

Family History.

Father aged 49.Health good.Ht.64 ins. Wt.146 lb.
Mother.d.aet.43. Cancer.Chorea. Ht 65 ins.Wt.142 lb.
Mother's uncle died of tuberculosis. The Wassermann
reaction of parents' blood was negative.
Siblings. Patient was first of three. One sister
was also microcephalic and died aged two years.
The other sister is normal.
There are no relatives of small stature.

Personal History.

Birth difficult.Forceps. Wt.7 lb. Walked,aet 10.
Never talked. Teething normal.
Illnesses.Chicken-pox,aet 10.

Physical Examination.

General Appearance. An apish looking boy with a fatuous grin. He has a very small head especially above the ears. Can only walk with assistance. Is doubly incontinent and cannot speak.

Height 56 ins. Symphysis-Vertex 27 ins. Symphysis-Soles 29 ins. Span 62 ins.

Cephalic Index 0.78. Greatest Circumference of head is 17 ins. Nasion-Occiput 9 ins. Binauricular 10 ins. Fat is normal. Posture is rather stooping and he walks with a scissors gait. Skin normal. Hair is very coarse and thick and the scalp is too big for the vault of the skull and is thrown into folds.

Mouth. Teeth are carious, but regular. No cleft palate. Eyes. Left internal strabismus. Fundi cannot be examined through patient's inco-operation.

Neck. No thyroid or cervical glands palpable.

Respiratory System. He is subject to bronchitis.

X-ray of chest does not show any gross abnormality.

Cardiovascular System. Pulse 94 per min. B.P. 130/90.

Heart not enlarged. Both sounds clear and closed.

The patient is very nervous and upset by examination.

Alimentary System. Abdomen is lean and lax. There is no enlargement of liver or spleen.

Urine contains neither albumin or sugar.

C.N.S. Pupils equal and active. Left internal strabismus. Other cranial nerves intact. Abdominal reflexes are absent. All deep reflexes are very much exaggerated but equal. The legs are affected worse than the arms. Plantar reflexes, extensor bilaterally. The patient suffers from spastic quadriparesis of

congenital origin.

Genitalia. These are normal. Both testes are in the scrotum and are of normal size and consistency. Seminal emissions occur. Pubic and axillary hair is normal in amount and distribution.

Laboratory Findings.

Wassermann Reaction negative.

Summary.

This microcephalic idiot shows the typical shape of head described before, where the reduction in size is confined to the head above the level of the ears. He shows a cephalic index of 0.78 in keeping with the usual findings, where the head is long and narrow, the longitudinal diameter being the least affected of any.

The mean of four cases of microcephaly in this group shows a stature of 57.75 ins. These patients tend to be small but they are not the smallest of defectives as a whole.

There is no sign, either in this patient or any of the others, that there is any delay in the development of the genital organs or secondary sexual characteristics,



Case No. 25. Example of Diffuse Hypertrophic
Amentia. Sex. Male. Grade Imbecile.
Age 21.

Family History.

The parents are both healthy and in no way abnormal. Siblings. He has a younger brother who is of a similar type but of higher grade and, therefore, is kept at home. Circumference of brother's head 23 in. Nearly all members of mother's side of the family have large heads although none is mentally defective and some are very intelligent. A nephew aged five already takes the largest size in school caps.

Personal History.

Birth was premature. Abnormality noticed at one year. Thought to be hydrocephalic. Teething was not delayed. Talked at two years and did not walk until he was three and a half years old. Never had fits.

Habits. A cheerful and well behaved imbecile.

Physical Examination.

General Appearance. A cheerful, young man with a large, massive looking head and a rather red face. Height. 66 ins. Weight 126 lb. Upper and lower measurements equal. Circumference of head 25 ins. Binauricular - $14\frac{1}{2}$ ins. Nasion-Occiput - $16\frac{1}{2}$ ins. Transverse Diameter $6\frac{1}{2}$ ins. Longitudinal - $8\frac{1}{2}$ ins. Cephalic index 0.79.

Fat. Well covered. Posture erect. Skin smooth and fine. Teeth strong, well calcified but crowded. The palate is very highly arched. Hair crisp and strong. Plentiful pubic and axillary hair and some growth on chest.

Congenital cataract of right eye. Palpebral fissures slope downwards and out. Sight in left eye seems normal. Left fundus normal.

No thyroid palpable. Genitalia mature.

No other abnormalities except right pupil smaller than left and both sluggish to light.

Summary.

A typical case of diffuse hypertrophic sclerotic amentia for comparison with nodular type. (No. 26)



Case No 26. Example of Nodular Sclerotic Amentia.

Sex. Male. Grade. Idiot. Age 27.

Family History.

The father is a patient in a mental hospital. He suffers from manic-depressive psychosis and is an epileptic.

The mother is in rather poor physical health and is unbalanced and extravagantly devoted to her idiot sons.

An elder brother was an idiot too but died aged 15.

Personal History.

Birth was premature (six months). Patient had infantile convulsions and is still epileptic. He had measles, pneumonia and bronchitis in infancy.

Always restless and excitable, even violent.

Facial rash noted since 1927.

Physical Examination.

General Appearance. He is quite a well built boy.

His face is covered with a red, papular rash.

Height 67 ins. Symphysis-Vertex 33 ins. Symphysis-Soles 34 ins. Span 66 ins. Weight 111 lb.

Fat. Well covered but not obese. Posture erect.

Skin shows hypertrophied tags all over the face and plaques on the forehead. The rest of the body is free of tags or cafe-au-lait patches. The skin is rather harsh and dry.

Teeth are quite well formed but are carious.

Hair is crisp and strong. Secondary sexual hair is normal in amount and distribution.

There are no gross abnormalities in any of the systems.

He is an epileptic and has two to four fits per month. He is completely dissociated mentally and stands all day grimacing and making stereotyped gestures. He is violent and spiteful if disturbed.

Summary.

A case of nodular sclerotic amentia. These are almost without exception epileptic. Epilepsy, mental deficiency and adenoma sebaceum are the three cardinal symptoms. Most of the other findings such as tumours in brain, kidney and heart, are usually only diagnosed post mortem. No signs of gross stature defect or sexual hypoplasia are found in these cases.

SUMMARY.

This investigation was carried out with a dual purpose in mind. First, to survey the subjects of Dwarfism and Infantilism generally, and to consider their relationship to mental deficiency in particular. Secondly, to show how unfavourably mental defectives as a class compare in stature with the general population.

Since dwarfism and infantilism are disorders of growth and development of a degenerative nature, one would expect to find them especially occurring amongst mental defectives, in whom other degenerative stigmata, both physical and mental, are very common. As may be seen from the illustrative cases given, the greater percentage of those types of dwarfism and infantilism which are noted in the general survey with which this thesis begins, are to be found in the group of 2,103 mental defectives in which this investigation was carried out.

The work begins with a consideration of the terms "Dwarfism" and "Infantilism", and although the definitions laid down by Meige (1895) and Levi (1910) in the case of dwarfism, and by Gardiner Hill (1937) in the case of infantilism, are accepted, it should be pointed out that there is much to be said in favour of postulating a statistically valid maximum height for male and female dwarfs in any given race.

This would mean that any individual whose height was less than the mean stature for his or her race and age by more than three times the standard deviation of that mean, could fairly and accurately be termed a dwarf. This standard would set the upper limit of dwarfism in British people at the figures of $59\frac{1}{2}$ inches and 51 inches, for males and females respectively.

A short historical section dealing with dwarfism is included mainly to show the types which were noted before the subject was broadened and complicated by the awakened interest of medical science and also, to draw attention to the fact that although dwarfism is no obstacle, in some cases, to success in various spheres of life, that success is limited to a few selected types such as the primordial, achondroplastic and rachitic.

The Racial and Developmental aspect of dwarfism is next considered and the similarity in the modes of production of dwarfed and infantile types, both in uncivilised and civilised peoples, is pointed out. From the racial point of view, stagnation, or deterioration, of development is often due to environment. This includes both climatic and nutritional factors. Those peoples who are of low development, and in whom development is stagnant, are to be found in the most unfavourable districts of the countries in which they live.

Within certain limits, the same applies to a race such as our own. Naturally, dwarfism is the

exception, rather than the rule, amongst the British, but in addition to the sporadic, or ontogenetic, types which form most of the examples in this country, we can find, in certain classes of the community, who dwell in the most unfavourable environment, a substratum showing stigmata of developmental regression and stagnation. This is what has been called "Developmental Infantilism", by Hastings Gilford, in contradistinction to "Evolutionary Infantilism" in more primitive races.

The basis of Developmental Infantilism is to be found amongst the members of a large group of mental defectives such as the one examined here. These people are drawn from one of the unfavourable districts in the country, the slums and industrial areas of Greater London, where the poor environment, with its accompaniment of malnutrition, overcrowding and disease, is responsible for a general deterioration of physique, and lowering of resistance, amongst its inhabitants. It is in this same environment that certain individuals with an inherent tendency to regress, and acted on by a series of predisposing factors, revert to a markedly inferior mental and physical type.

Gilford points out that environment may be external, internal, or both. This gives rise to two types of developmental infantilism: an adaptative type in response to external noxious factors such as specific fevers, alcohol, chronic infections and syphilis, and a correlative type in response to a degen-

degenerated/
system or organ, the body "correlating" itself to
the degenerated part.

Whether or not we need to draw this close distinction, the fact remains that mental defectives are the product of a set of noxious factors such as syphilis, alcohol, tuberculosis and epilepsy, working hand in glove with an unfavourable environment.

At this stage we can see the two lines of enquiry which we are pursuing, emerging from the facts above. On the one hand we have a general deterioration of physique amongst the mental defectives who form the lowest stratum of this society, and on the other hand we have the production, amongst those defectives, of numerous dwarfed and infantile types in whom, evidently, the tendency to developmental regression is more marked than in their fellows.

In order to provide a comprehensive table of the types of Dwarfism and Infantilism met with in mental deficiency practice, the opinions and classifications which a series of authors have given over the past thirty years, have been examined, and the various views expressed by them are shown in the discussions which follow their tables of classification.

From these discussions, and from a collation of the types dealt with therein, a table was drawn up, (Table IX), in which all the well authenticated forms of dwarfism and infantilism which one would expect to find in mental deficiency practice, are placed. In addition to this table, 26 illustrative cases are presented, which show most of the

types of dwarfism and infantilism included in Table IX, and all of which have been discovered in this colony of mental defectives.

If we examine Table IX we find that, although naturally enough, the broad outlines of the table are applicable to dwarfism and infantilism in general, in dealing with mental deficiency we have certain variations and discrepancies from current opinions to consider, and an important addition to make.

Although, in view of the advance of endocrinological research during the past thirty years, the endocrine basis of dwarfism and infantilism is now all-important, it must be remembered that the clear cut "syndrome" is comparatively rare and the primary cause of dwarfism and infantilism often obscure. It seems to be the case, in mental deficiency, that the primary cause is often non-endocrine, and very often neurological, the endocrines being involved in a widespread regressive change secondary to the neurological (or other) cause.

However much emphasis we place, then, on the important endocrine types which we know of, there must be equal space given to types whose causes are essentially non-endocrine. Some of these are quite well defined, such as achondroplasia, spinal caries and rickets, in the case of dwarfism, and coeliac and renal disease, in the case of infantilism. But together with these, we have a large number of dwarfed and infantile subjects, amongst mental defectives,

whose condition seems consequent on neurological trauma or disease, and another appreciable group whose condition, although probably ultimately due to endocrine failure, is primarily caused by multiple aetiological factors such as have been outlined earlier in giving Gilford's views on Developmental Infantilism.

Thus, in spite of the progress of the past thirty years, this table will still contain types which are covered by Gilford's hypotheses. Such are Simple Cachectic Infantilism (Gardiner Hill 1937) and Infantilism due to Traumatic Brain Disease with or without Spinal and Limb Deformity.

The condition of Mongolian Imbecility has been added to those types which exhibit dwarfism alone. It was not found in any of the tables examined, although Tidy (1939) mentions it apart from the table which he gives. As is shown in the special section devoted to the subject, Mongols are all of diminutive stature and their average height falls within the limits of dwarfism. This colony contains 55 Mongolian Imbeciles, of whom 35 are males. In none of those males were there any signs of infantilism and puberty was rarely even delayed. None of them had undescended testes, and function, in so far as it could be assessed from erectile power and masturbatory activities, was not interfered with. Amongst the female Mongols only three were completely amenorrhoeic and menstruation, in general, was no more irregular than in

other types of defect. Any irregularities that do occur seem to be due to the accompanying hypothyroidism so often seen in these cases and replacement therapy improves menstrual irregularity.

There is thus no foundation for any statement that infantilism is a regular feature amongst Mongolian Imbeciles although the condition is still said to occur by some authorities.

Dwarfism due to Congenital Syphilis has been excluded from Table IX because it was definitely established, that in this group, stature was not affected enough by this disease to warrant its inclusion. 4.1 per cent of the patients here suffer from congenital syphilis and this appears to be about the average figure for large groups of defectives. It was actually found that both sexes were above the average height for the group.

These, then, are the principal alterations made in the table as applied especially to mental defectives. As the dwarfism due to Mongolian Imbecility seems to have been neglected in most of the tables, and as Congenital Syphilis, a common cause of defect, seems to have but an ill-established claim to dwarfing in the condition, the examination of other well known types of mental deficiency was undertaken to determine whether or not they led to an appreciable degree of stunting and infantilism.

What are called Primary Aments form 80 per cent of the total number of defectives. The cause of Primary Amentia, which is endogenous, is still unknown and the label "germinal defect" is

still attached to it. The majority of defectives thus fall into this Primary Amentia class and it comprises a group in whom physical stigmata are many and varied. The average stature of Primary Aments naturally corresponds closely to the average for defectives as a whole.

About 10 per cent of these primary aments, however, fall into several comparatively small groups in each of which some distinct aetiological factor, or factors, is at work to produce a close resemblance between the patients in each group. Such are the Mongols, microcephalics, sclerotic aments, oxycephalics, naevoid aments and hypertelorisms.

The stature of Mongols has already been discussed and the last three are so rare that no cases of oxycephaly or hypertelorism were found in this group, and only two examples of naevoid amentia, both of whom were of average height.

From the small numbers of true microcephalics found, it was deduced that the stature was less than the average defectives but was, at times, by no means dwarfed, as in the case of one female microcephalic who was 0.04 inches taller than the average for the group. Four adult, male microcephalics were, on the average, $1\frac{1}{2}$ inches taller than the Mongolian Imbeciles. These findings are in disagreement with Tredgold (1939) who states that microcephalics, as a class, are the smallest of defectives. Adult, male microcephalics were not found to be infantile although puberty was sometimes delayed.

The other subdivision of Primary Amentia.

examined was the class of Sclerotic Aments. These patients form two sub-groups. The best known is the Nodular Type, often called Epiloia, Adenoma Sebaceum or Tuberous sclerosis. This colony, at one time, contained 25 cases of this rather rare condition but, as the patients tend to die young, only six could be assessed. These were all females as the males were mostly adolescent. The average height of these six female cases of epiloia was 57.75 inches, which is 1.21 inches below the average for the group as a whole. The condition does not lead to dwarfing and, far from being infantile, male adolescents present, rather, signs suggestive of macrogenitosomia!

Diffuse sclerotic amentia is even more uncommon and in the only case seen here, as in his brother who lives at home, stature and sexual development were not unduly interfered with, both being above the average for the group.

The remaining 20 per cent of mental defectives is made up of Secondary Aments in whom the mental deficiency is secondary to an exogenous cause. As has been pointed out earlier in this discussion, neurological lesions are responsible for a great number of the low grade types in the group and these cases form two important groups in Table IX.

In the first, the cases are dwarfed by extensive spinal and limb deformity following on lesions such as birth trauma and congenital mal-developmental paralyses such as Little's Disease. Stature in these cases is merely dwarfed by the

twisting of the spine and contracture of the limbs, and this may be so extensive as to make actual measurement difficult.

It would seem from the cases examined that extensive involvement of the pyramidal system leads to stunted growth and great delay in the process of sexual maturation. The case illustrative of Cachectic Infantilism, shown in this series, demonstrates to what a degree this may extend when pyramidal involvement, and a growth period beset with illness, are combined.

The second group contains patients who owe their deficiency to brain trauma caused by such conditions as epidemic encephalitis, hydrocephalus, cerebral syphilis, amaurotic family idiocy and epilepsy. Epidemic encephalitis, being the cause of a special "behaviour disorder" type of mental defect which does not manifest itself necessarily until adolescence, does not give rise to any constant degree of stunting. No conclusive evidence about stature could be given from the 11 cases examined, the males being 3.2 inches taller and the females being 2.86 inches shorter than the group average.

On the other hand, epidemic encephalitis is responsible for a marked upset in the sex-life of the subject and menstrual irregularities, or even amenorrhoea, are common. This is thought to be due to involvement of the pituitary and hypothalamic areas, as water and carbohydrate metabolism is upset and obesity common, especially in females.

Hydrocephalics, although smaller on the

average, than other defectives, do not exhibit marked dwarfing and all the male cases were over five feet in height. Although basal meningitis, of acute or syphilitic type, is a frequent cause of hydrocephalus this does not seem to lead to primary or regressive sex disorders, as seen in the encephalitics.

Congenital Syphilis as a cause of dwarfism, has been excluded, in this group at any rate, earlier in the discussion. When, however, the central nervous system is involved and juvenile paralysis of the insane ensues together with pyramidal involvement and consequent physical paralysis, then growth is greatly stunted and sexual characters remain infantile. These patients then enter the Cachectic Infantilism group. In those adult females in whom the central nervous system was not involved, menstruation was almost as regular as in all the other female defectives. Figures are shown in the section devoted to this subject.

Degenerative brain changes, such as are seen in Amaurotic Family Idiocy, lead invariably to a condition of cachectic infantilism and dwarfism but, more rarely, the disease may arise in adolescence, and in this juvenile type the stature and sex life may have proceeded normally until the onset of the disease. Thereafter, both will soon become totally arrested.

Epilepsy, which is included in the degenerative group, is the commonest accompaniment of mental defect. 604 patients; 28.7 per cent of this group, suffer from epilepsy. In order to assess the

effect, if any, of epilepsy on stature, only so-called "idiopathic" epilepsy can be considered, as epilepsy is such a common accompaniment of diseases which in themselves would tend to stunt the stature. 103 male, able-bodied epileptics who showed no other nervous lesions, were measured. Their intelligence quotients ranged from below 25% to 70% so that all grades of defect were included. The average stature was 63.76 inches, which is 1.46 inches above the average for the group. None of these showed any gross degree of sexual retardation. Idiopathic epilepsy does not, therefore, give rise to any dwarfing or infantilism.

Having estimated the height and assessed the incidence of infantilism in the individual groups of mental deficiency, we pass on to compare the stature of mental defectives as a whole with that of the normal population and with their fellows in the U.S.A.

Tredgold (1937), quoting the investigation of the British Association in 1882, shows that lunatics and aments are amongst the smallest classes of the population. As the defectives in this colony nearly all come from the London district, a comparison was made between them and a middle-class London group measured by Pearson and Lee. (1903). The figures show that there is an enormous difference. Male defectives are 5.8 inches, and female defectives 3.52 inches shorter than the normals. Even when we compare the stature of defectives with that of the poorer artisan class measured by Dukes (1905), there is still a marked bias in favour of the latter, actually 4.2 inches,

in males. Dukes does not include females in his group.

The average height of male defectives in this group was found to be 62.3 inches, and that of female defectives, 58.96 inches. These figures were found to correlate very closely in the case of females, with the figures given by Goddard (1912) for American defectives. (Table XIV). American male defectives were, however, over two inches taller.

In order to compare the figures of different groups accurately, the predominant grades of defect in each of the groups must be known, and we do not know whether Goddard's group may not have contained larger numbers of feeble-minded males than this one, which is predominantly imbecile.

The importance of eliminating this tendency to error, even in large groups, is shown by the final investigation carried out here, where the stature in each grade of mental deficiency is shown. Only adult, able-bodied, simple aments are measured and, from the figures, it can be seen that although stature does not increase steadily in proportion to the rise of intelligence quotient yet, if we divide a large group of defectives into three grades, as is empirically done for mental testing and statutory legal returns, then there is a significant difference in the heights of the idiots, (I.Q. below 25%), the imbeciles, (I.Q. 25-50%) and the feeble-minded, (I.Q. 50-70%), taken as groups, and there is the most marked difference between able-bodied feeble-minded patients and able-bodied idiots.

Idiots, as a group, include a large percentage who are crippled, and whose stature is reduced by various physical causes which we have examined, so that, in comparing their stature with that of the highest, feeble-minded grade, we must measure only able-bodied members of each type, the sole difference between members of each grade being one of intelligence alone. The results, shown in Tables XVI and XVII, demonstrate the progressive stepping up of stature in each grade from idiot to feeble-minded.

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The salient points and conclusions of this summary are shown overleaf.

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CONCLUSIONS.

1. It would be useful to lay down a statistically valid maximum for the height of dwarfs in any given race. This might be a value equal to the mean stature for the race and age minus three times the standard deviation of that mean.

2. This would give a value for English dwarfs, of $59\frac{1}{2}$ inches for males and 51 inches for females.

3. Mental Defectives have many points in common with the Developmental Type of Infantilism postulated by Hastings Gilford in 1911, and the condition of mental deficiency owes much to the same biological factors.

4. In mental deficiency, equal emphasis should be placed on endocrine and non-endocrine types of dwarfism and infantilism, and many of the cases have a primary neurological basis, endocrine involvement being secondary to widespread regression.

5. Mongolian Imbecility should be included in tables of dwarfism and infantilism, and especially in any classification devoted to mental deficiency.

6. Dwarfism due to Congenital Syphilis is not the rule in mental deficiency although 4-5% of patients suffer from this disease. Only cases who show involvement of the central nervous system, exhibit any marked signs of stunting and infantilism, and these may be included in the Simple Cachectic Infantilism group.

7. Microcephalics, although stunted more than their fellow defectives, are not the smallest class of defective.

8. Epiloia (sclerotic amentia) does not lead to dwarfing and infantilism.

9. Spinal and limb deformity, consequent on brain trauma, either congenital or acquired, is responsible for many cases of dwarfism in defect. If pyramidal involvement is severe, then it is often accompanied by generalised infantilism.

10. Epidemic encephalitis causes no appreciable stature change, but gives rise to a marked menstrual irregularity and appears to exert its influence by involvement of the pituitary and hypothalamic areas, disturbances of water, carbohydrate and fat metabolism being common.

11. Hydrocephalics are stunted, mostly owing to limb paralyses and deformities. Infantilism is not usually seen.

12. Amaurotic Family Idiocy causes generalised cachectic infantilism, but if it arises later, in adolescence, growth and sexual development are normal up to the onset.

13. Idiopathic epilepsy has no affect on stature or sexual development.

14. Mental Defectives compare most unfavourably with the normal population, in stature, even when compared with the working classes.

15. There is little discrepancy between the average heights of English and American female defectives. Males are two inches taller in U.S.A.

16. The height of mental defectives rises significantly through the three grades of idiocy, imbecility and feeble-mindedness. Lack of intelligence alone, seems to produce a smaller type. Able-bodied male idiots are 5.9 inches shorter on the average than able-bodied feeble-minded patients.

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